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*ORIGINAL STUDY*

# Primary bone and soft tissue tumours : epidemiological data from a non-referral teaching department in Belgium

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**Primary bone and soft tissue tumours are rare in a non-referral teaching department. The incidence varies greatly among the different subtypes and every Orthopaedic surgeon will encounter one or more benign or malignant lesions during their careers. History, clinical examination and technical investigations are of great importance, but basic knowledge and basic principles are necessary for a correct clinical practice. It was the purpose of this study to raise awareness towards such pathology by exemplifying our cases from a two-year period in our community-based hospital and if necessary, to refer patients to specialised sarcoma centres.**

**Keywords** : Primary bone and soft tissue tumours ; non-referral department ; epidemiology.

## INTRODUCTION

The diagnosis of primary bone and soft tissue tumours is rare in a non-referral teaching department. Most orthopaedic surgeons are not familiar with this heterogeneous group of tumours and many do not know how to handle such lesions. The majority of primary bone and soft tissue tumours follow a benign clinical course. Primary bone neoplasms are extremely rare, accounting for only 0.2% of human tumours *(16)*. Sarcomas on the other hand are a heterogeneous group of malignant neoplasms arising from mesenchymal cells. They can occur in almost any anatomic site and are defined by diverse histopathologies of the affected organ, tissue, bone or cartilage. The RARECARE-project, estimated an incidence of soft-tissue sarcoma and bone sarcoma of 4.7/100,000 and 0.8/ 100,000 respectively with the incidence of STS slightly higher in Northern Europe compared to Eastern Europe *(19,20)* tissue, bone or cartilage. Due to their heterogeneity, estimates of sarcoma incidence, prevalence and survival are rare. We estimated the burden of sarcoma in Germany from a large unselected cohort of patients from routine healthcare. Methods : We utilized the AOK PLUS health services research database covering complete medical information on 2,615,865 individuals from the German federal state of Saxony from 2005 to 2012. Persons were defined as sarcoma cases if they had -4 medical accounts with respective ICD-10 code C49 (soft-tissue sarcoma This article reviews the spectrum of tumours collected during a 2-year period in our department. We were interested in what a group of fifteen Orthopaedic surgeons might encounter in their daily practices and how we managed this less familiar part of our orthopaedic specialty. The aim

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is to increase the awareness of the treating physician on the diagnosis and management of Orthopaedic-related tumours.

## PATIENTS AND METHODS

AZ Delta Roeselare is a supra-regional orthopaedic department with 15 consultants active in all fields of orthopaedics and trauma. We have over 70.000 patient contacts in the outpatient clinic and perform just over 10.000 orthopaedic and trauma surgeries each year. Some of our staff members are Ph. D. and are part-time affiliated with university departments.

All staff members and residents were asked to report bony and soft-tissue sarcoma as well as any bump, lump or mass during a two-year period from August 2016 until August 2018. We excluded lipomas, ganglion cysts, mucoid cysts, epidermal inclusion cysts, warts, skin carcinomas and gout tophus. Indeterminate histopathology at the moment of biopsy or surgery was checked retrospectively. We collected data of 75 patients ; 69 benign tumours and 6 malignant tumours were diagnosed.

## RESULTS

Seventy-five persons met the inclusion criteria of which 69 (92%) were benign tumours and 6 (8%) were malignant tumours. Figure 1 shows the anatomical location of the tumours. Of the benign tumours, the upper limb (n=40) was more affected compared to the lower limb (n=26). The hand (n=31) was more commonly involved than the other regions. Localized PVNS (n=11), followed by enchondroma (n=5) were the main diagnoses. Regarding lower limb involvement, the femur was the preferred site for 11 tumours with osteochondroma (n=3) and NOF (n=3) comprising just over half the cases. For the benign group, more females (n=40) than males (n=29) were affected. Another pattern was seen for the malignant group, where more males (n=5) than females (n=1) were affected. Mean age in both groups was 40 years old. Eighteen different benign

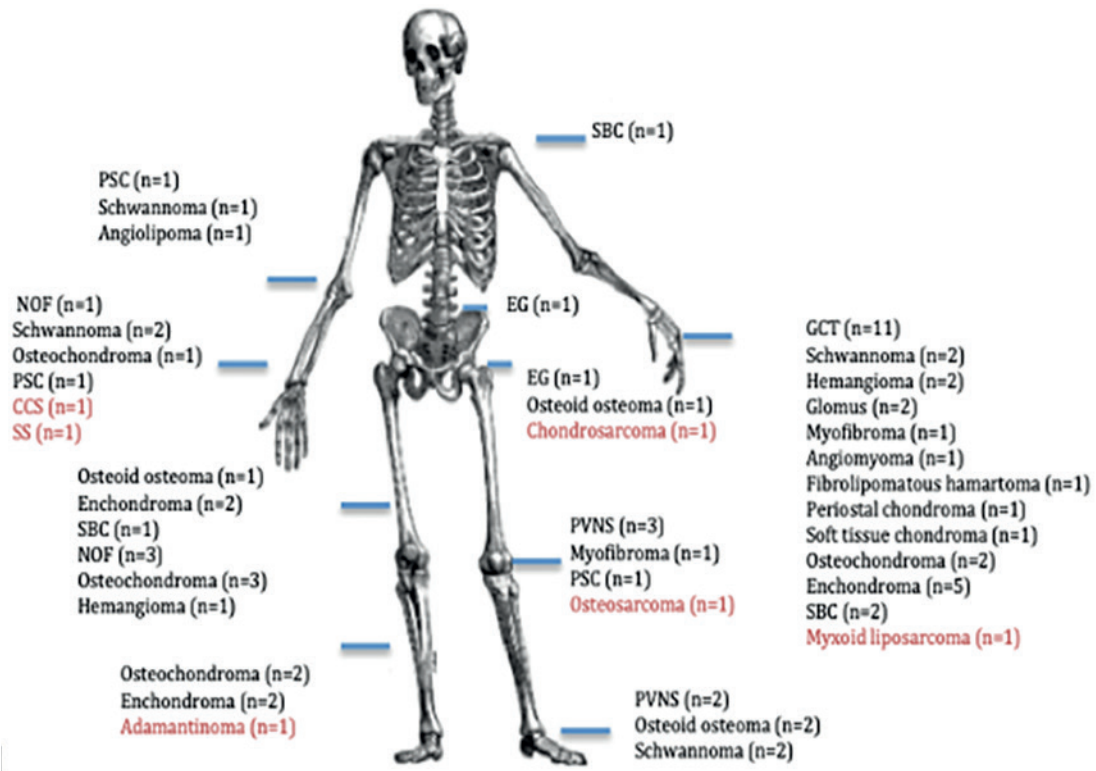


***Fig 1.*** — Anatomical distribution of benign (black) and malignant (red) tumours across the body.

lesions and 6 different malignant lesions were found and they are briefly discussed hereafter.

*Localized PVNS or localized Giant Cell Tumour L-GCT* was the most common primary soft tissue tumour and accounted for 12 cases (33%). Eleven tumours were seen at the hand and only one at the foot. There was an equal sex distribution and no hand predominance or location preference was seen. On the left hand the following digits were affected ; DII (n=3), DIII (n=2) and DV (n=1) while on the right hand ; DI (n=1), DII (n=1), DIV (n=3) and DV (n=1) lesion. Preoperative diagnoses were nine times GCT, one trigger finger, one Dupuytren nodule and the one at the toe was unclear. All patients received preoperative sonography and only the GCT of the foot was staged with an MRI. All lesions but one were resected. Histopathology confirmed the GCT diagnosis and up until now, no recurrence was seen.

Four patients were diagnosed with *diffuse-type Giant Cell Tumour or D-GCT* of which three occurred about the knee and one was found about the ankle. MRI confirmed all diagnoses and the PVNS were arthroscopically resected in all knees. Only one knee needed an extra posteromedial open excision. The PVNS inside the ankle joint was diagnosed on MRI and then referred to a specialised tumour centre.

*Schwannoma* was diagnosed in seven patients. Five tumours were located in the upper limb and two in the lower limb. All upper limb schwannomas were preoperatively evaluated with sonography and the two ankle schwannomas were staged with MRI. All but one schwannomas were excised and histopathology confirmed the diagnosis.

Three patients presented with *primary synovial chondromatosis (PSC).* Three different locations were seen ; the elbow, the wrist and the knee. Diagnoses were made according to the location. The knee was preoperatively evaluated with MRI and after multidisciplinary consultation, arthroscopic excision was proposed because of progressive increasing pain. The upper limb tumours were diagnosed with X-ray and MRI. Conservative treatment was the treatment of choice.

*Glomus tumour* only occurred twice and was localised at the subungual region of the fourth digit on the right hand and at the second digit on the left hand. Diagnosis was made clinically and both tumours were excised. Histopathology confirmed the diagnosis and no recurrence was seen.

Three *haemangiomas* were diagnosed of which two occurred at the left hand and one originated at the left vastus lateralis. The hand tumours were preoperatively diagnosed with sonography and the suspected haemangioma at the thigh was staged with X-ray, sonography and MRI. All three haemangiomas were excised and histopathology confirmed capillary haemangioma.

*Enchondroma* was the most common benign bone tumour and accounted for nine cases. They were predominantly (n=5) seen in the hand and both femur and tibia accounted for 2 enchondromas each. These lower limb tumours underwent follow-up with MRI and no adverse effects were noted. Located at the hand, two pathologic fractures were treated conservatively with good results after healing. Because of a history of a contralateral pathologic fracture, curettage and bone grafting was the preferred treatment for one enchondroma

*Osteochondroma* was the second most common benign bone tumour and reported in 8 patients. This tumour was predominantly (n=5) seen in the lower limb. Three of them occurred at the femoral site of which two osteochondromas underwent follow-up with MRI and only one was resected. One of the two osteochondromas at the tibial site was resected because of posterior tibial tendon irritation. One tumour at the ulna was being investigated with MRI because of the potential accelerated growth at the age of thirteen. Two osteochondromas occur-red in the hand of which one was resected and one was referred to a specialised tumour centre because of indistinct features seen on X-ray, sono-graphy, MRI and scintigraphy. The definitive diagno-sis was a “Nora-lesion” or Bizarre Parosteal Osteo-chondromatous Proliferation (BPOP).

Four *Non-Ossifying Fibromas (NOF)* were dis-covered as incidentalomas of which three occur-red in the distal femur and one occurred at the radius.

Four *simple bone cysts (SBC)* presented at our department of which two were situated at the wrist and both the proximal femur and proximal humerus were involved once. The cysts in the lunate and scaphoid were resected and grafted with

good results. The simple bone cyst in the proximal humerus was complicated with a fracture. Healing was uneventful after conservative treatment. One simple bone cyst was situated in the proximal femur compromising the mechanical stability. Treatment consisted of resection, bone grafting and application of DHS, yielding a good result.

*Osteoid osteoma* accounted for four cases all occurring at the lower limb. At the acetabulum, a fracture preceded the diagnosis of an osteoid osteoma for which CT-guided radiofrequency was proposed after healing of the fracture. The same treatment was proposed for the calcaneal tumour. For the talar osteoid osteoma a resection was performed with good results. The aforementioned tumours were all preoperatively staged with X-ray, CT and scintigraphy. In only one case the diagnosis was delayed due to a concomitant impingement problem requiring arthroscopic hip surgery. Because of persistent postoperative pain at night an additional arthro-CT of the hip was carried out, revealing the diagnosis of an osteoid osteoma. Also in this case, CT-guided radiofrequency was proposed.

*Eosinophilic granuloma (EG),* which is also known as Langerhanscelhistiocytosis, accounted for two cases. In one case the tumour presented at the right pedicle of S1 in a five-year old boy. The diagnosis was made based on X-ray, MRI, CT and scintigraphy and treatment consisted of resection of the tumour with good results. The other EG was found in the left acetabulum. The patient was referred to a specialised tumour centre because the results seen on different technical investigations were inconclusive. After one year, spontaneous regression was noted and the patient did well.

Six benign tumours were encountered only once. A *soft tissue chondroma* at the left second digit was excised after appropriate preoperative investigation with sonography and radiography. One *benign angiomyoma* at the first webspace was diagnosed by sonography and treated by surgical excision. A *fibrolipomatous hamartoma* at the median nerve was excised after two years of persistent pain. The diagnosis of a *periostal chondroma* at the right fifth digit was made after microscopic investigation because preoperative sonography, X-ray and MRI were inconclusive. One *angiolipoma* at the left elbow was excised after being diagnosed on pre-operative sonography. And finally, *one myofibroma* was located around the knee, requiring surgical excision. Preoperative diagnosis was made on sonography and MRI.

Six patients (8%) were diagnosed with a malignant tumour during a two-year period. The upper limb was most commonly affected. Three malignancies (50%) were found at the wrist, while the lower limb and the acetabulum accounted for the other half of the cases.

One *adamantinoma* in a 22-year old woman was already treated more than ten times with wide excision surgery in Algeria. She presented with a severely MRSA-infected leg with complete resorption of distal tibia and fibula. She was admitted to our hospital and given IV antibiotics. A transtibial amputation was the only treatment possible.

The diagnosis of one *clear cell sarcoma* was made in a 41-year old male presenting with an ulnar volar swelling of the right wrist. Medical history and clinical examination were normal. Sonography and MRI confirmed a ganglion cyst. Excision was done and the histopathology report confirmed a nodular tenosynovitis. Recurrence of this mass occurred one year later and sonography reported a giant cell tumour. A re-excision was performed and the histopathology report showed a clear cell sarcoma. Further staging was done after multidisciplinary consultation and the patient was referred to a specialised tumour centre.

A fifteen-year history of ulnar dorsal wrist pain preceded the diagnosis of one *synovial sarcoma* in a 34-year old patient. Further additional medical history and clinical examination were normal. MRI did not reveal a clear diagnosis and surgical excision biopsy was planned. The histopathology report showed a synovial sarcoma and excision margins were not free of tumour cells. Further staging was done after multidisciplinary consultation and the patient was referred to a specialised tumour centre.

One *osteosarcoma* was encountered in a 24-year old male presenting with pain of the left knee. Clinical examination showed limping and a swollen knee. Urgent MRI and CT showed signs of an osteo-sarcoma. Immediate referral to a specialised tumour centre for further staging was performed.

The diagnosis of a *chondrosarcoma* was made in a 50-year old male presenting to the Emergency department after a fall from a height. X-ray showed a fracture of the anterior wall of the acetabulum and a CT scan showed an osteolytic lesion expanding to the left iliac wing. Differential diagnosis consisted of fibrous dysplasia and chondrogenic tumour. Staging with MRI assumed a chondrosarcoma, confirmed by the oncologist in a specialised tumour centre. The patient was then immediately referred to a specialised tumour centre for further staging and treatment.

One *myxoid liposarcoma* was diagnosed in a 50-year old male with persistent swelling of the left hypothenar region. No additional details in the medical history and on clinical examination. Sonography showed a benign nodular structure and MRI reported a ganglion cyst. Excision was performed and the histopathology reports from three independent University hospitals differed according to malignancy but they agreed on an atypical presentation of a myxoid tumour. Up until now, no recurrence was noted.

## DISCUSSION

*Tenosynovial giant cell tumours (TGCT),* formerly known as pigmented villonodular synovitis (PVNS) are rare, benign tumours, arising in synovial lining of joints, tendon sheaths, or bursae.

Two types are distinguished : localized, either digits or extremity, and diffuse lesions. The incidence varies greatly and depends on the form with an annual incidence rate of TGCT ranging from 1.8 per million to 50 per million. As in our study, the incidence of localized TGCT (including digits) is about four times more than the diffuse form of TGCT. They affect generally young patients below the age of 40 years with an equal sex distribution. The age in our study group is 49,5 years old. The diffuse-type TGCT is a more destructive and locally aggressive tumour of which 75% are located around the knee joint. In our patients, no recurrence was seen but the lifetime recurrence risk is up to 15% for L-TGCT and up to 55% for D-TGCT *(12,9)*.

Schwann cells across the nerve can give rise to *Schwannomas*, also known as neurilemmomas.

They represent less than 5% of soft-tissue neoplasms of the upper extremities and they usually occur in patients aged 30 to 60 years and have no race or sex predilection. As in our study group, there is a higher incidence in the flexor surface of the upper limb, since the concentration of nerve fibres is higher over that region and the upper extremities are twice as likely to be involved as the lower limbs *(15,21)*.

*Primary synovial chondromatosis (PSC)* is a rare condition characterized by the formation of multiple cartilaginous nodules in the synovium of joints, tendons, and bursae. It is thought to be a self-limiting metaplasia but a relative risk of 5% for malignant change was observed. The average age at presentation is 41 years with more males than females affected, as opposed to our group of which the age averages 60 years old and only females were affected. It is described in literature that the most common site is the knee joint (70%) followed by the hip (20%) *(6)*.

*Glomus* bodies are thermoregulatory shunts concentrated in the dermis of the fingertips and other peripheral sites subject to excessive cold. They present with a triad of localized tenderness, severe paroxysmal pain and sensitivity to cold. We found two cases but they tend to be under-recognized. One study described that an average of 2.5 physicians, including psychiatrists, were consulted before the correct diagnosis was made and that the duration of symptoms averaged 10 years *(18)*.

*Haemangiomas* are common benign soft tissue tumours, comprising 7-10% of all soft tissue tumours. They occur before the age of 30, but there is no consensus for sex predilection. Cutaneous haemangiomas are the most common and we also found two originating at the hand. Intramuscular haemangiomas are sometimes perceived as a sporting injury. They are more common in the lower limbs with the quadriceps the most common intramuscular site as it was with the vastus lateralis in our group. Haemangiomas enlarge slowly and their growth may be accelerated with a growth spurt but they can also spontaneously regress and malignant transformation is rare *(17)*.

*Non-ossifying fibroma* belong to the most common focal lesions in bones. NOF do not cause clinical

symptoms and are usually incidentally found in X-ray examinations performed because of an injury, which is in accordance with our findings. Incidence is estimated in up to 30% of the asymptomatic population in the first and second decade of life. Similar to osteocartilaginous exostoses, they develop in the metaphysis, in the region of intensive bone growth *(4)*.

*Simple bone cysts (SBC)* are benign fluid filled cavities that enlarge over time, resulting in thinning of the bone. Usually these cysts are reported in the metaphyseal areas of long bones with open physes. 85% of UBCs occur almost exclusively in children and adolescents. The reported peak is between the ages of 3 and 14 years with the mean age at diagnosis being approximately 9 years. They occur twice as much in boys as compared to girls *(14)*. These data are consistent with those in our study but we also report one SBC at the lunate and one at the scaphoid in 2 males of older age.

*Osteoid osteoma* is the third most common biopsy analyzed benign bone tumor after osteochondroma and NOF. It occurs predominantly in long bones of the appendicular skeleton with the lower extremity being more affected than the upper extremity. Commonly long bones particularly the femur and tibia are involved, followed far behind by bones of the feet, with a predilection for the talar neck. CT is the modality of choice for the diagnosis, as it was made clear in our CAM impingement-case. Most osteoid osteomas undergo spontaneous healing. Surgical treatment is an option for patients with severe pain and those not responding to NSAIDs *(13)*.

*Eosinophilic granuloma (EG)* is a rare, benign tumor-like disorder that is the most common manifestation of Langerhans-cell histiocytosis (60-80 % cases), accounting for less than 1 % of all bone tumours and mostly (80 %) children and adolescents. We had one case where the tumour was situated in the lumbar region and one in the acetabulum. Reviewing the literature, EG preferably are situated in the thoracic spine and account for 6.5-25 % of all spinal bone tumours *(1)*.

*Adamantinomas* are low-grade malignant neoplasms with a predilection for the diaphysis of the tibia. These tumours are extremely rare, accounting for < 1% of primary bone tumours, commonly affecting male patients between the ages of 20 to 50 years. Adamantinomas are usually treated with wide-local excision, but historically by an amputation *(10)*. In our case, amputation was needed because of MRSA infection with complete resorption of distal tibia and fibula.

*Clear cell sarcoma (CCS)* is an extremely rare (1%) and aggressive subtype of sarcoma thought to derive from the neural crest cells, most often observed in the third decade, without any gender predilection. As opposed to our case at the ulnar volar wrist site, CCS arises in 95% of the cases in the lower extremities, particularly the foot and ankle. It presents as an indolent, growing and painless mass situated in the deep soft tissue or beneath the fascia. The malignant nature of the swelling is seldom suspected as we also had to perform re-excision *(11,7)*. This was not different in the patient in our series where a second procedure was indicated after obtaining the histological report.

*Synovial sarcoma (SS)* is a malignant mesenchymal tumour. It is most common among children and adults, between 10-35 years old. In our series the tumour was located at the ulnar dorsal aspect of the wrist while review of the literature shows that the incidence at the lower limb is twice that of the upper limb *(3)*.

*Osteosarcoma* derives from primitive bone-forming mesenchymal cells and is the most common primary bone malignancy. It commonly occurs in the long bones of the extremities in the metaphyseal area. Osteosarcoma has a bimodal age distribution, having the first peak in the 10-14-year-old age group and the second peak in adults older than 65 years of age. Males (5,4 per million persons) are more affected than females (4,0 per million persons). As it is in our case, the most common sites are around the knee, the distal femur (42%), the proximal tibia (19%) and the proximal humerus (10%). *(5)*while malignant bone tumors make up just over 10% of sarcomas. The risks for sarcoma are not well-understood. We evaluated the existing literature on the epidemiology and etiology of sarcoma. Risks for sarcoma development can be divided into environmental exposures, genetic susceptibility, and an interaction between the two. HIV-positive

individuals are at an increased risk for Kaposi's sarcoma, even though HHV8 is the causative virus. Radiation exposure from radiotherapy has been strongly associated with secondary sarcoma development in certain cancer patients. In fact, the risk of malignant bone tumors increases as the cumulative dose of radiation to the bone increases (p for trend <0.001

*Chondrosarcoma* is a malignant tumor, producing a cartilaginous matrix without tumor osteoid. Its estimated annual incidence is 1 in 200,000 and rises with increasing age. Its etiology is unknown ; chondrosarcoma may arise de novo in normal bone or may undergo malignant transformation from a previously benign cartilaginous tumor *(2)*.

*Liposarcomas* are the second most common type of soft tissue sarcomas, 30-50% of these are of myxoid subtype. They are a genetically distinct variant of liposarcoma, characterized by a t(12:16) translocation *(8)*.

Reviewing the literature, the epidemiologic data are fairly consistent with our small group of patients over a two-year period.

Limitations of the current study are the small number of subjects and the selection bias. The two co-authors are specialized in hand surgery and were more aware of reporting data of patients presenting with any bump, lump or mass.

## CONCLUSION

During our daily practice the incidence of bone- and soft tissue tumours is rare but every Orthopaedic surgeon will encounter one or more benign or malignant lesions during their careers. Correct clinical practice is of utmost importance to prevent 'whoops'-procedures or re-resections. Histological diagnosis of these lesions represents a challenge for pathologists, sometimes requiring need for expert pathology review. Outcome is dictated by a raised awareness towards such pathology and community based hospitals should implement clinical practice guidelines and refer patients to specialised sarcoma centres when needed.

## REFERENCES


1. **Angelini A, Mavrogenis AF, Rimondi E, et al**. Current concepts for the diagnosis and management of eosinophilic granuloma of bone. *J Orthop Traumatol* 2017 ; 18 : 83-90.
2. **Arshi A, Sharim J, Park DY, *et al***. HHS Public Access 2018 ; 42 : 644-652.
3. **Asian S, Kuruva SP, Bala S, et al**. Clinicopathological features, treatment and survival outcomes of synovial sarcoma 2018 ; 7 : 270-272.
4. **Błaż M, Palczewski P, Świątkowski J, *et al***. Cortical fibrous defects and non-ossifying fibromas in children and young adults : The analysis of radiological features in 28 cases and a review of literature 2011 ; 76 : 32-39.
5. **Burningham Z, Hashibe M, Spector L, *et al***. The Epidemiology of Sarcoma. *Clin Sarcoma Res* 2012 ; 2 : 14.
6. **Davis RI, Hamilton A, Biggart JD**. Primary Synovial Chondromatosis : A Clinicopathologic Review and Assessment of Malignant Potential 1998 : 683-688.
7. **Deenik W, Mooi WJ, Ph D, *et al***. Clear Cell Sarcoma ( Malignant Melanoma ) of Soft Parts A Clinicopathologic Study of 30 Cases 1965.
8. **Dürr HR, Rauh J, Baur-melnyk A, *et al***. Myxoid liposarcoma : local relapse and metastatic pattern in 43 patients 2018 : 1-7.
9. **Ehrenstein V, Andersen SL, Qazi I, *et al***. Tenosynovial Giant Cell Tumor : Incidence , Prevalence , Patient Characteristics , and Recurrence . A Registry-based Cohort Study in Denmark 2017.
10. **Houdek MT, Wenger DE, Sherman CE, *et al***. Adamantinoma of bone : Long-term follow- up of 46 consecutive patients 2018 : 1-5.
11. **Ibrahim RM, Jensen SS, Juel J**. Clear cell sarcoma-A review. *J Orthop* 2018 ; 15 : 963-966.
12. **Mastboom MJL, Verspoor FGM, Verschoor AJ, *et al***. Higher incidence rates than previously known in tenosynovial giant cell tumors Higher incidence rates than previously known in tenosynovial giant cell tumors A nationwide study in The Netherlands 2017 ; 3674.
13. **Noordin S, Allana S, Hilal K, *et al***. Osteoid osteoma : Contemporary management 2018 ; 10.
14. **Noordin S, Allana S, Umer M, *et al***. AC. *Ann Med Surg* 2018.
15. **Ozdemir O, Kurt C, Coskunol E, *et al***. Schwannomas of the hand and wrist : long-term results and review of the literature 2005 ; 13 : 267-272.
16. **Perera JR, Pollock R**. Management of benign bone tumours. *Orthop Trauma* 2017 ; 31 : 151-160.
17. **Report C**. Intramuscular haemangioma of the thigh in a basketball player 2004 : 346-348.
18. **Stewart DR, Sloan JL, Yao L, *et al***. Diagnosis, management, and complications of glomus tumors of the digits in neurofibromatosis type 1 2010 ; 47 : 525-532.
19. **Stiller CA, Trama A, Serraino D, *et al***. Descriptive epidemiology of sarcomas in Europe: Report from the RARECARE project. *Eur J Cancer* 2013 ; 49 : 684-695.

**20**. **Trautmann F, Schuler M, Schmitt J**. Burden of soft-tissue and bone sarcoma in routine care. Estimation of incidence, prevalence and survival for health services research. *Cancer Epidemiol* 2015 ; 39 : 440-446.

**21**. **Yuk C, Tang K, Fung B, *et al***. Schwannoma in the Upper Limbs 2013 ; 2013.