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CASE REPORT

Axillary fossa tumours in children : rare and easily missed

Sandeep SHEWALE, Roderick DUNCAN, Dermot MURPHY, Milind RONGHE

From the Royal Hospital for Sick Children, Glasgow, Scotland

Three children presenting with pain in the arm and variable neurological signs are described. Each child had several hospital visits before being accurately diagnosed with a soft-tissue sarcoma in the axillary fossa. All presented with increasing pain at rest or on shoulder movement, and with some evidence of neurological dysfunction in the upper limb. Two of the girls had unremitting night pain.

Delays in diagnosis of sarcomas are common despite internationally agreed referral guidelines, which are readily accessible. In children with unusual upper limb neurological symptoms, it is essential to have a high index of suspicion, and examine the arm including the axilla, thoroughly, and refer them early for a specialist opinion.

Keywords : axillary fossa ; soft-tissue tumours ; children ; delayed diagnosis.

INTRODUCTION

Three cases of children presenting with pain in the arm and variable neurological signs are described. Each child had several visits to a local Accident and Emergency department and to other specialities before being accurately diagnosed with a soft-tissue sarcoma of the axillary fossa. All eventually required treatment with multimodal therapy including chemotherapy, radiotherapy and surgery.

CASE REPORTS

Case 1

A girl aged 2 years and 2 months was brought to the Accident and Emergency in our institution, her parents having found a mass in the axilla. She had presented to her local hospital on three different occasions during the previous three months, with pain on abduction of her arm, and reluctance to use her left arm. Each time she was investigated with radiographs of her shoulder which were reported to be normal. A detailed physical and neurological examination did not appear to have been done.

■ Sandeep Shewale, Consultant Orthopaedic Surgeon.
Monklands Hospital, Airdrie, Scotland.
■ Roderick Duncan, Consultant Orthopaedic Surgeon.
■ Dermot Murphy, Consultant Oncologist.
■ Milind Ronghe, Consultant Oncologist.
Royal Hospital for Sick Children, Glasgow, Scotland.
Correspondence : Sandeep B. Shewale, 27, Kirkdene Avenue, Newton Mearns, Glasgow, G77 5RN, Scotland.
E-mail : shewalesandeep@yahoo.com

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On first presentation to our institution, she had a palpable mass of 8×6 centimetres in the anterior axillary fold. Her pain had become unremitting and severe. Neurological examination revealed difficulty in full extension of fingers and inability to oppose the thumb. She had no sensory loss.

Diagnostic and staging investigations showed this to be an undifferentiated soft tissue sarcoma arising from the pectoral muscle and invading the brachial plexus, with no lung metastasis.

Case 2

An 8 ½ year-old right handed girl was referred by her General Practitioner and was seen by the paediatricians at her local hospital with a 6-month history of intermittent right arm pain, spasm and progressive weakness and reduced sensation of her arm and hand. She was found to have a moist and slightly warmer right hand, marked limitation of shoulder abduction, full painfree neck movement and no palpable axillary and supraclavicular mass. A provisional diagnosis of brachial plexus neuropathy (post viral infection) was made and she was referred to the neurologists at another tertiary level hospital. An MRI of the neck was carried out, which did not include the brachial plexus. She was reviewed by the paediatricians four months later and there were no signs of improvement. Her pain had become constant and the neurological deficit profound. She had become left handed. A month later she presented to her local Accident and Emergency department as her mother had noticed a lump in the girl's axilla, which the girl herself had noticed about 2 weeks prior. She was promptly referred to our institution.

On presenting to us, she had a large palpable mass in the right axilla, displacing the scapula posteriorly. She had profound weakness and sensory loss in the entire arm below C5, with no Horner's syndrome. Radiographs suggested a soft tissue tumour and showed moulding of the proximal ribs. MRI showed a $12 \times 9 \times 11$ cm mass arising from the lateral anterior chest wall, extending to the proximal humeral shaft and involving the Brachial plexus. Biopsy of the lesion showed a soft tissue Ewing's sarcoma. Surgical resection involved shoulder disarticulation.

Case 3

A 2-year-old girl first presented to her local Accident and Emergency department with a painful left arm and discomfort in the hand of about 2 months duration. She also had had a minor fall a week prior to her visit. After taking radiographs of her wrist, a plaster splint was applied for a wrist sprain. The pain worsened and became constant, the child routinely crying with it at night. Her mother also noticed that she complained of pain when being lifted with hands in her axillae. She made several visits to the Accident and Emergency over the following six months. Her parents however brought her to our institution due to the severe, unresolving pain.

On examination she was noted to be reluctant to abduct her left shoulder due to pain. Her parents had noted her reluctance to use her left hand while playing. There was an asymmetry in the upper arm, with a 5 cm palpable mass in her left axilla. Power in the left arm and grip strength was reduced significantly. Radiographs were normal.

MRI showed a 7×3 cm ovoid mass around the medial aspect of the left upper arm, close to the neurovascular bundle but not involving the bone. Diagnosis after incisional biopsy was that of a malignant peripheral nerve sheath tumour with rhabdosarcomatous differentiation (Triton tumour). Staging CT scan of her lung showed the presence of bilateral lung metastasis.

During surgery the tumour was found to be arising from the median nerve. Surgery involved enbloc excision including the median nerve.

DISCUSSION

All three of these children presented with increasing pain at rest or on shoulder movement, and with some evidence of neurological dysfunction in the upper limb. Two of the girls had unremitting night pain. In children, the differential diagnosis of upper limb pain at rest and neurological deficit is either brachial plexus neuritis or a malignant tumour (*4,6,7*). The presence of a mass on or near a major nerve makes the diagnosis of a malignant nerve tumour more likely. There are obvious

problems in assessing the neurological function in young children, particularly for those clinicians in whom this is not a regular part of their practice. A specialist opinion should be obtained at an early stage, as delay in diagnosis is first of all, likely to mean that more extensive surgery will be necessary, and secondly, it may influence survival (*1,9*).

Primary tumours of the brachial plexus are unusual (*3*), but account for 20% of all peripheral nerve tumours (*8*). Malignant tumours have a predilection to arise from major nerves especially the brachial plexus, as compared to benign tumours (*6*). Whenever there is discordance between clinical signs and cervical Imaging, other conditions should be considered and thoroughly investigated (*2*). In case 2, no further examination of the axilla is documented after the initial visit and the MRI did not include the brachial plexus. MRI has a major role in the characterising and staging of tumours of the brachial plexus (7). In spite of major advances in MRI, differentiating benign and malignant tumours can be difficult, and correlation with clinical symptoms is of diagnostic value (*6,7*).

Delays in diagnosis of all sarcomas are common (*1,9*) despite internationally agreed referral guidelines, which are readily accessible : www.dh.gov.uk/cancer ; www.show.scot.nhs.uk. Examination of the axilla should be an integral part of the upper limb examination but is not routinely taught as part of the examination of either the musculoskeletal system or nervous system (*5*). The purpose of this report is to alert health professionals who see a child with unusual limb symptoms to the possibility of an axillary tumour, to encourage them to have a high index of suspicion, and examine the arm including the axilla, thoroughly, and refer them early for a specialist opinion.

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