Brachial Plexus' Schwannoma in a Child

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Abstract

Children's schwannoma is a rare condition, generally occurring in a sporadic way. Its aetiology is still not fully understood. We report the case of a 10-year old girl who presented a left shoulder mass, along motility reduction of the left upper limb for 24 months before presentation at our service. A biopsy resection of the mass gave the diagnosis and 6 months after surgical resection, no complication occurred.

Keywords: Case report, children's schwannoma, peripheral nerve tumors

INTRODUCTION

Peripheral nerve tumours (PNTs) are rare conditions. These lesions can be reactive or malignant or benign. Among the latter, two are most frequent in the paediatric population: neurofibroma and schwannoma (neurilemoma). While the first is linked with neurofibromatosis type 1, the second is usually sporadic.^[1]

We report a case in a 10-year-old girl. We discuss the frequency of schwannoma in the paediatric population, its diagnosis, treatment, histology and outcomes.

CASE REPORT

A 10-year-old girl was referred to our service for the management of a left shoulder mass. History reported the beginning at 24 months by a progressively growing mass inducing limited mobility of the left upper limb, for which, 4 months later, the patient has been consulted at a paediatric surgical service in Dakar, where a left shoulder X-ray and magnetic resonance imaging (MRI) have been done. For technical reasons, the patient was referred to us.

We noted a left suprascapular globular hard and painful mass, extending to the lateral part of the left supraclavicular fossa. It was adherent to the underlying tissues. Realised X-rays did not show any skeletal alteration MRI done a year ago reported a solid mass with septa, located in soft tissues anterior to

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the scapula [Figure 1]. Due to financial constraints, a new look MRI could not be obtained. The patient underwent a biopsy-resection whose exploration showed that the tumour invaded the scapulohumeral articulation's capsule, muscles of the rotator cuff and extended up to the midline. The tumour was adherent to the brachial plexus (BP) and blood vessels. Resection was done in four parts, with laborious dissection of the BP. The four parts of the resected mass weighted 400 g. The largest measured 11 cm \times 7 cm \times 6 cm [Figure 2].

Macroscopical examination revealed superposable whitish tissues, in a fascicular disposition. Microscopical examination showed fusocellular proliferation composed of Schwann cell's, containing wavy low-grade nuclei with verocay bodies. Cells were arranged in crisscross fasces, storiform in a fibrillar matrix containing focal oedema and being denser and hyalinised. There were <3 mitosis and the tumour was encapsulated. The conclusion was a Grade 1 schwannoma according to the French National Federation of Cancer Centres.

On the post-operative (PO) day 1, the patient suffered anaemia (7.2 g/dL) which has been corrected. She has been discharged on day 7 postoperatively. We reviewed her 15 days later. She had no complaint and has already been partially reusing her left thoracic limb. Six months later, our patient has no complaint. She uses her left upper limb with a discrete

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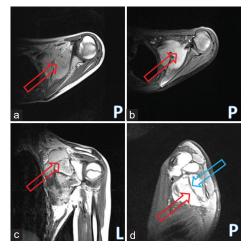


Figure 1: Left shoulder's magnetic resonance imaging. On horizontal section a mass anterior to the scapula with low intensity signal in T1-weighted (a) and high intensity signal in T2-weighted (b). It measured 628 cm \times 127 cm \times 90 mm and had septa within it as shown on the sagittal (c) and coronal (d) sections

weakness. There was no recurrence of the mass on the left shoulder and sensitive neurological examination was normal.

DISCUSSION

Schwannoma is a benign PNT (BPNT) arising from Schwann cells. In the paediatric population, schwannoma is the second-most common BPNT.^[2,3] It represents 5% of all soft-tissue neurogenic tumours in children.^[4] Most occur in vestibular location as non-vestibular schwannomas (NVSs) represent 1.2% of paediatric schwannoma. Most NVSs are paraspinal, in the head and neck or in limbs.^[5] In our patient, it occurred in the BP, which is considered in the upper extremity. As stated, it is one of the common sites of NVSs. It equally occurs in males and females.^[5]

Schwannoma usually presents as an isolated firm mass on the trajectory of a nerve, mobile perpendicularly to the suspected nerve course. Pain is frequent and motor or sensory deficits are rare.^[6,7] Physical examination should check for clinical manifestation of associated syndromes such as NF2, schwannomatosis, Carney complex or dominant syndrome.^[1,4,7] Our patient did not present any of the features which could refer to one of the cited syndromes.

In our patient, we performed MRI, which along with computed tomography are the best major imaging of schwannomas. On MRI, the tumour has iso-intense or intermediate images in T1-weighted while it shows uniformly hyperintense images in T2-weighted. MRI has the possibility to depict the tumour and its capsule, as well as the affected.^[1,6] For the reported case, MRI could unfortunately not precise the origin of the tumour. This can be attributed to the important size of the mass.

Surgical resection is the gold standard of schwannoma's treatment. Optimally, enucleation along with preservation of



Figure 2: Macroscopic aspect of the mass after surgical resection. Note its globular appearance as well as its well-defined limits

the affected nerve should be the preferred excisional mode.^[4] This consists of opening the tumour's capsule, enucleate it and section, under magnification, the fascicle from which the tumour originates.^[2,8] If the neurological deficit is severe, extended excision is justified.^[6] In our patient, we performed an endocapsular enucleation and preserved the BP.

Schwannoma has a macroscopic appearance of a well-limited encapsulated mass. Through its section, an eccentric fashion can be macroscopically seen,^[1] this was the case in our patient. Microscopically, schwannomas are characterised by a biphasic pattern composed of dense and cellular areas (Antoni A), alternating with less dense and hypocellular areas (Antoni B). Within Antoni A areas, cells may palisade to form verocay bodies. The proportion of Antoni A and Antoni B areas highly varies.^[1,3,5] In our case, histological findings showed a predominant Antoni A tumour. Immunohistochemistry for S-100 protein results in high distribution and intensity of staining.^[3] This has not been done in our case due to financial constraints.

Schwannomas have a good prognosis. PO numbness, weakness and pain syndromes resolve progressively, but pain and the neurologic deficit will persist in case of nerve resection.^[8,9] The recurrence rate is <5%.^[4] Our patient presents a good outcome, without any recurrence sign 6 months after surgery.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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