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Axillary mass in pediatric age: rare case of schwannoma of the median nerve

Massa ascellare in età pediatrica: raro caso di schwannoma del nervo mediano

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Summary

A 15-year-old male patient came to our attention for the appearance of a large mass in the right axilla for about 4 months. In the anamnesis, a first surgical stage of open exploration is reported, which disproved the suspicion of lymphadenopathy. Following preparatory investigations, a diagnosis of suspected schwannoma of the median nerve was attributed, confirmed operatively and by histological examination. The post-operative follow-up was uneventful.

Key words: neuroma, pediatric neoformation, pediatric benign tumours, Schwann cell, neurolemmocytes

Riassunto

Un paziente maschio di 15 anni è giunto alla nostra attenzione per la comparsa di una grossa massa in regione ascellare destra da circa 4 mesi. In anamnesi viene riportato un primo approccio chirurgico di esplorazione open, che ha permesso di escludere il sospetto di linfoadenopatia. A seguito di indagini di approfondimento è stata posta diagnosi di sospetto schwannoma del nervo mediano, confermata operativamente e dall'esame istologico. Il follow-up post-operatorio è stato regolare.

Parole chiave: *neurinoma, neoformazioni pediatriche, tumori benigni pediatrici, cellula di Schwann, neurolemmocita*

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Introduction

Axillary fossa masses can be due to a very different kind of medical causes like lymphadenopathy, lymphomas, metastasis, benign tumors. Schwannoma is a benign encapsulated tumor that grows in the peripheral nervous system. It develops in the nerve sheath from Schwann cells, the principal glia of the peripheral nervous system. These tumors are more common in people between 20-50 years old, with no difference about sex or race [1 2]. These usually appear as isolated lesions even

though sometimes they can be related to neurofibromatosis type 2 and arise in multiple clusters [3]. Malignant transformation is rare [4]. They are found mostly in the head and in the neck (25%) or in the flexion surfaces of legs and arms, only for the 5% in the axillary fossa [5]. It could take many years from the presentation to diagnose the mass. Because of its rarity, some cases can be missed or misdiagnosed, so it should be kept in consideration as one of the differential diagnosis about an axillary mass originating from a nerve branch [6].
In this report we describe an axillary mass, initially misdiagnosed as an axillary inflamed lymph node, that eventually resulted in a median nerve schwannoma in a teenaged patient.

Case report

A 15-year-old male patient presented to his primary care physician with a palpable mass in his right axillary fossa since 4 months. No pain or other symptoms were referred. He had no fever, 2 kilos weight lost after a period of diet. Hematological exams were normal. Axillary ultrasound shown a lymphadenopathy of the axillary fossa with a node of about 30 mm and an altered echo structure (Fig. 1).
A pediatric oncohematologist visited the patient and confirmed the diagnosis of reactive lymphadenopathy. She suggested a pediatric surgical evaluation to consider the lymph node removal and analysis.
The patient underwent surgery after two weeks: during surgical exploration a well vascularized and roundish mass was identified, characterized by a proximal and a distal peduncle. Trying to dissect the vascular and nerval structures, surgeons reported the flexion of the third finger of the right hand. Due to the suspect of a wrong diagnosis, it was decided to stop the procedure and to add other diagnostic studies. The skin was sutured and the operation postponed. Patient parents were informed and agreed (Fig. 2).
After surgery the patient was investigated with a right axillary fossa MRI and brain/encephalic trunk MRI to investigate

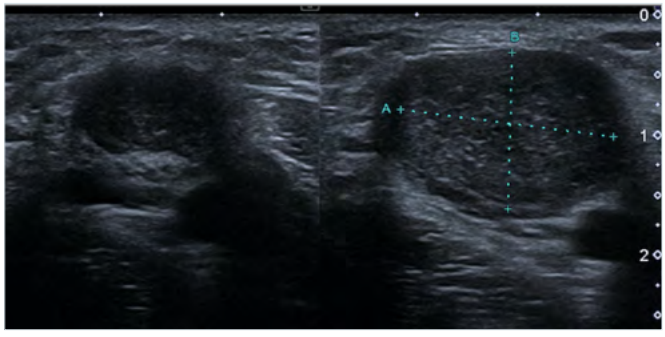

Figure 1. Sonografic appearence of axillary mass at the first examination.

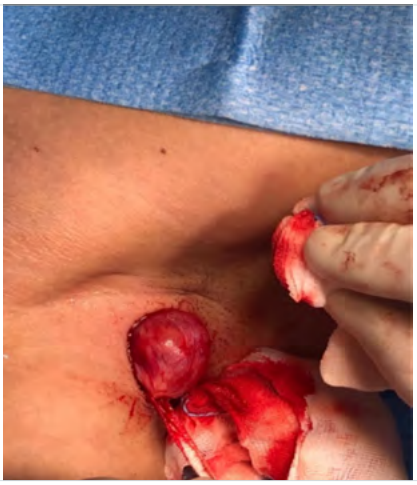

Figure 2. First surgical stage: isolation of the mass with proximal and distal peduncle.

further nature of the neoformation and verify the eventual presence of any tumors. MRI refers: "near to axillar vessels, there's a roundish formation with sharp edges, 23 x 19 mm on the axial plan, 26 mm on craniocaudal axis. According to clinical-anamnestic profile the image indicates the suspect of neurinoma. No other alterations were found in brain or in the encephalic trunk.". The patient had a second sonographic examination performed by a specialist to investigate the ultrasound aspect of schwannoma. He was investigated with an EMG, no alteration in conduction were reported.
After this finding, the Hand Surgery and Microsurgery Unit took charge of the patient.
The pre-operative evaluation consisted of anamnesis, physical examination of the tumor, superficial sensory function testing (touch, pain, and static and dynamic sensory discrimination), muscle tone, and strength examination as well as testing for Hoffmann-Tinel sign. We managed to test the relation about the mass and the median nerve, so with flexion-extension movement of the elbow and the wrist (median nerve neurodynamic stretching) we could see the mass moving in the axillary fossa. The patient reported about some kind of sporadic and paroxysmal dysesthesia and paresthesia in the median nerve territory that he had never mentioned before.
The patient underwent a second surgery in general anesthesia (Figs. 3, 4). The incision was made directly on the longest axis of the mass. The vessels were detected and protected; accurate hemostasis permitted to recognize the structures involved. We isolated proximally and distally the nerve branch and we protected the peduncles with surgical loops. The perineurium has been incised keeping attention to its preservation. We dissected the adherent fascicles from the proximal pole of the tumor to the distal one. Once they were completely detached and the mass isolated, it was excised

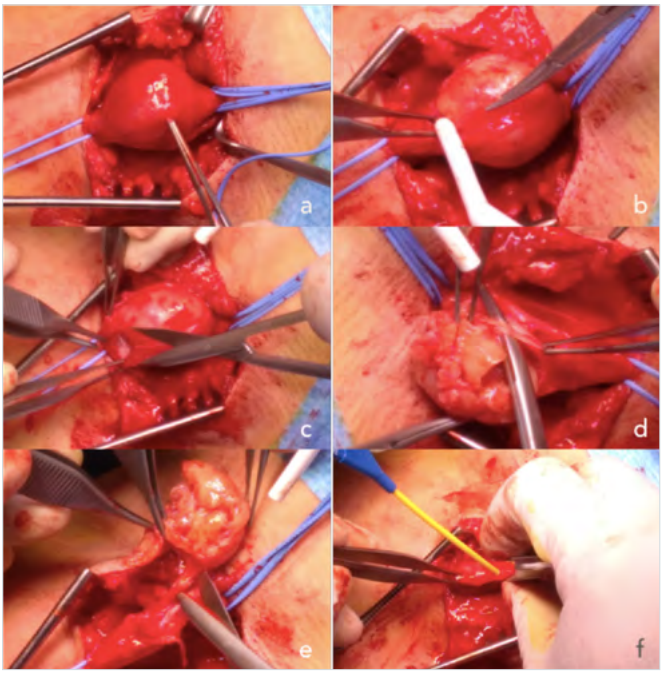

Figure 3. Second surgical stage. a) Median nerve isolation; b) Epineurium incision. c) Schwannoma's capsule incision. d) Fascicula isolation. e) Schwannoma excision. f) Human fibrin glue application.

and sent for the histopathological examination. The nerve fascicles of the median branch were left intact but stretched and adapted to the capsuled mass; this macroscopic aspect oriented for a possible Schwannoma. We stitch the epineurium and we applied human fibrin glue (Tisseel® - Baxter). The skin was closed with stitches, a soft bandage was made as medication. We allowed the patient to freely move the arm after 2 days since surgery. The scar has been treated with dedicate cream application for 2 months after surgery.
Histopathology confirmed the diagnosis of right axillary fossa schwannoma of the median nerve. Its section showed a tumor formed of benign-looking spindle cells with hypocellular areas. Immuno-histo-chemistry was positive for S100 and BC34 with no evidence of malignancy.
The postsurgical progress was regular, some mild neurological sensitive symptoms concerning the middle finger endured for the next three months after surgery and they were treated with neurotrophic (Palmitoilethanolamide, PEA) integration. These symptoms completely disappeared at the 3 moths post-op clinical evaluation.

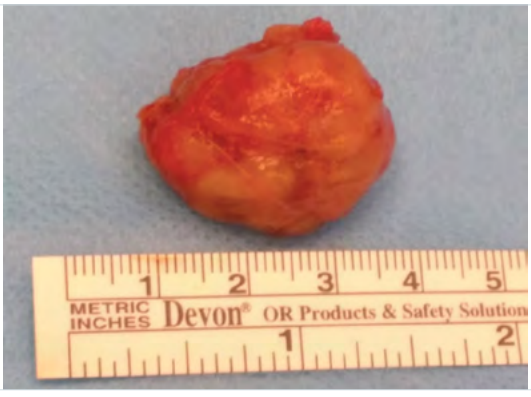

Figure 4. Excised schwannoma.

Discussion

A lymph node abscess incision, biopsy or excision, in pediatric age, are usually suggested by oncohematologists or by pediatricians for common infections or in cases of antibiotics therapy unresponsive masses.
Generally, neck, inguinal and axillary area are the most common surgical sites.
In pediatric age, without suspicious of neoplastic disease, ultrasound is the most common radiological test used; on ultrasound a normal lymph node has a hypoechoic cortex but often shows a central hyperechoic hilum containing fat and intranodal blood vessels referred to as a hilar line. The hilar line is more prominent in adolescents.
Many different pathologic conditions can present as axillary masses in addition to lymph node enlargement (fibroadenomas, hamartomas, fat necrosis, carcinomas, lipomas, schwannomas). Awareness of the variety of these disease entities and characteristic sonographic findings can aid in correct diagnosis of an axillary mass [7].
Verocay described Schwannoma in 1908 as a slow-growing benign tumor of the sheath of peripheral nerves [8]. It is the most common peripheral nerve tumor and it affects mainly the head, the neck and the extremities. Axillary schwannomas are very uncommon as described in Gosk et al.: just six cases of axillary schwannoma between all the patients treated for extremities schwannomas between 1985 and 2013 were found [9] we present our experience in operative management of schwannomas and analyse results of treatment. Clinical material consisted of 34 patients, in whom 44 schwannomas located in extremities were excised between 1985 and 2013. Thirty-five tumours originated from major peripheral nerves and 9 from small nerve branches. Post-operatively, in the first group of tumours, pain resolved in 100%, paresthesias in 83.3%, and Hoffmann-Tinel sign in 91.6% of the patients. Improvement in motor function was noted in 28.5% of the cases, in sensory function: complete in 70%, and partial in 15%. The most frequently affected major peripheral nerves were the ulnar (11 tumours). Only 10% of schwannomas are diagnosed in patients less than 21 yo [10].

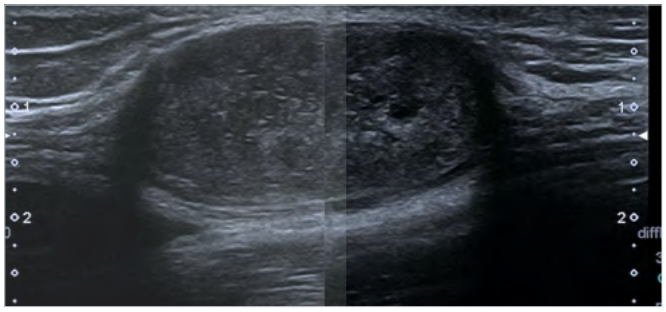

Figure 5. Sonografic imaging of schwannoma associated to median nerve.

In this case, the patient presented is a 15yo young male without any risk factors for developing schwannoma during pediatric age. One of these is schwannomatosis[3] which increases the potentiality of developing multiple schwannomas during the second decade of life.
We cannot find other cases about an axillary median nerve schwannoma in a pediatric patient referred in literature on the main scientific platform research.
We suggest to test the mass with upper limb and surrounding joints active and passive movements. If the mass moves in association with the stretching of the nerves it could indicate a nerve origin mass. Pain, Tinel test, muscular deficit or sensitive disturbs have to be considered.
The imaging evaluations of axillary mass should start with US examination. Distinction of a nerve sheath tumor from abnormal lymph nodes may be difficult because abnormal lymph nodes could lose their hila and can exhibit posterior acoustic enhancement. However, the most reliable method to differentiate a lymph node from a nerve sheath tumor is the identification of the associated nerve [11] (Fig. 5).
Schwannoma sonography shows a well-defined oval homogeneous hypoechoic mass with or without posterior enhancement [12]. Collagen deposit areas appear as a coarse echo texture or as focally increased echogenic areas. An echogenic ring within the mass (target sign) is rare but a pathognomonic feature of nerve sheath tumors, as in particular for long aged schwannomas [13]. An echogenic capsule is usually seen, and cystic spaces represent the presence of degenerated portions of the mass. Sonography can provide confirmation of a neurovascular bundle adjacent to the mass [14]. In addition, it could be useful performing a sonoelastography in order to evaluate the elasticity of the lesion and to provide more information about its eventual growth pattern [15].
Diagnosis and classification of the tumor is challenging. Fine needle aspiration (FNA) could be a method even though it is difficult to recognize tissue architecture pattern [16]. A trucut [17] biopsy is another option for diagnosis as shown in a reported case of gastric schwannoma diagnosed using this

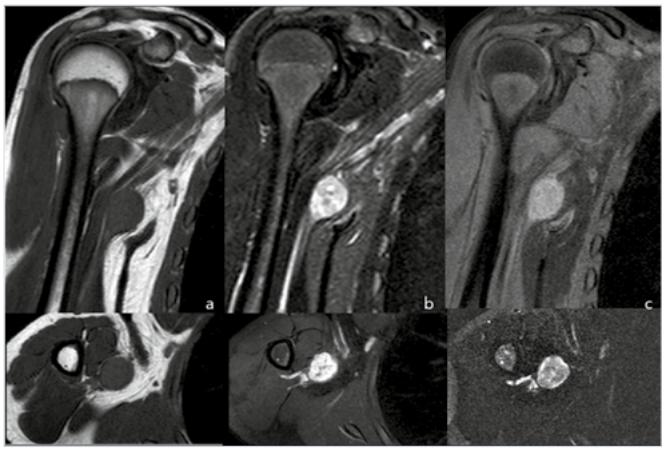

Figure 6. MRI appearance of axillary schwannoma of the patient. a) T1-weighted. b) T2-weighted. c) STIR.

procedure [18]. A new study published in the early 2020 by Roberto J. Perez Roman et al. reported that benign tumors (namely schwannoma and neurofibroma) biopsy poses an unacceptably high risk for neurological deficit [19], changing the usual approach to the diagnosis.
The MRI can surely provide to visualize the anatomy of brachial plexus and the interrelated anatomic structures on relatively artifact-free images with a large field-of-view, characterized by better detection and characterization of the lesions. This is the gold standard for differential diagnosis. On MRI [20], the tumor appears as a well-defined mass of intermediate signal intensity on T1-weighted images and as a mass of high intensity on T2-weighted images, with an inhomogeneous central low-signal area and strong enhancement after contrast agent administration. On T2-weighted images, the peripheral hyperintense signal is due to the presence of myxoid tissue, and the central low signal intensity is due to the presence of fibrocollagenous tissue [21] [22] (Fig. 6).
The treatment for schwannoma of the brachial plexus is resection of the tumor [23]. The tumor is located eccentrically rising from few fascicula in the peripheral nerve and is well encapsulated by the perineurium. These fascicula are often without functional meaning because of the tumor. For this reason, it has generally been accepted that careful dissection under magnification could achieve complete enucleation without causing further neurological deficit [24]. Different surgical techniques are described. In any case surgeons should attempt to perform a gross total resection of benign peripheral nerve sheath tumors without causing new neurological deficits. An ideal dissection plane can be identified between the pseudocapsule and true capsule [25]. Muramatsu et al. in their study show the advantages of the effective intracapsular enucleation using the microsurgery technique instead of extracapsular [26].

Conclusion

Axillary schwannoma is a very challenging condition itself, in terms of diagnosis and treatment. In particular, it becomes more difficult in pediatric patients. Lymphadenopathy is of course the first option in case of axillary mass in young patients but as we saw it could not be the only possible diagnosis. Because of unspecific symptoms, in many cases it could be mismanaged. It is very important to perform an accurate clinical exam, including Tinel sign and evaluating the relative motion of the mass correlated with the movement of the arm. US examination of the axilla fossa is the first imaging exam but often it is not enough to identify the right nature of the mass. Once made the definitive diagnosis, the surgeon can perform and choose the best way to approach and remove the mass.

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