

# CHILDREN'S FIBROUS HAMARTOMA: EXTENSIVE INJURY INVOLVING BRACHIAL PLEXUS

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## SUMMARY

**Objective:** To present a case of fibrous hamartoma in a late-dried infant presenting as an extensive injury, involving vascular and neural elements of brachial plexus. **Methods:** Clinical case study and pertinent literature review. **Results:** Male child, with right axillary irregular tumoral mass, of which onset occurred at 2 months of age and related to BCG vaccine application, being treated with anti-tuberculosis agents, not responding to therapy. Upon biopsy, the injury was diagnosed as children's fibrous hamartoma, and, after a fast growing period, was submitted to total surgical exeresis.

**Conclusion:** The juvenile fibrous hamartoma is a rare benign tumor, typically occurring within the first year of life in boys, most commonly located at axillary gap. The differential diagnosis is performed with soft parts tumors in general, and, in right axillary location cases, with axillary adenopathies caused by responses to BCG vaccine. Treatment approach is total exeresis of the injury and the prognosis is favorable.

**Keywords:** Hamartoma; BCG Vaccine; Fibroma. Neoplasms, Connective Tissue

## INTRODUCTION

Children's fibrous hamartoma is a rare tumor, with only about 200 cases reported in literature, the absolute majority of those with a diameter < 4 cm, typically axillary or thoracic. Here, we describe one case of a large tumor, involving vascular-nervous structures of the brachial plexus after a fast growth period in an infant, discussing the differential diagnosis and treatment, with a review of related literature. Patient care was provided in accordance to the standards accepted by the Committee on Ethics of the University-Hospital Antonio Pedro.

## MATERIALS AND METHODS

Case study. We have searched in literature the keywords 'infantile fibrous hamartoma' on MEDLINE, EMBASE and LILACS databases.

## RESULTS

A white, 2-month old male infant was taken to the primary healthcare center presenting with a 2-cm superficial hard tumor, and low range of motion at his right axilla. The lesion was attributed to a lymphangitis secondary to BCG vaccine and treated with isoniazide for three months.

Due to a continued increase of the nodule, two months later, the infant was referred for evaluation at the pediatric surgery service at University-Hospital Antônio Pedro, being performed an incisional biopsy, which revealed an infantile fibrous hamartoma. A fast lesion growth was seen along arm's

medial and longitudinal axis, reaching 15 cm of longitudinal diameter at the age of 9 months (Figure 1) and causing a difficult motion to the child as an effect of the mass, despite of a normal neurological test. Requested imaging tests (US, CT and NMR) evidenced an anteromedial mass on right arm adhered to humerus and involving nervous and vascular structures (Figure 2). Total resection of the lesion was then planned and performed in conjunction, by disciplines of orthopaedics and pediatric surgery, after an informed consent given by parents, considering the risks of vascular and neural injuries. During surgical procedure, a yellowish hard tumor, with undetermined borders, no capsule, infiltrate-like at muscle planes, and involving the whole brachial plexus, brachial artery, and basilic and brachial vessels was found. The lesion was adhered to the humeral proximal third, with no invasion or bone injury. It was fully dried after the isolation of the brachial artery and ulnar, median, and radial nerves, which remained intact (Figure 3), with no vascular or neurological complications. Axillary vein ligation was required. We used an antithrombotic prophylaxis during the first 5 postoperative days with low molecular weight heparin, evolving with no interurrences. Postoperative histological test confirmed the diagnosis of children's fibrous hamartoma.

## DISCUSSION

The infantile fibrous hamartoma was described by Reye<sup>(1)</sup> in 1956 (childhood's fibromatous subdermal tumor), and less than 200 cases have been reported by global literature so far, with 8 being described in Latin-American lit-

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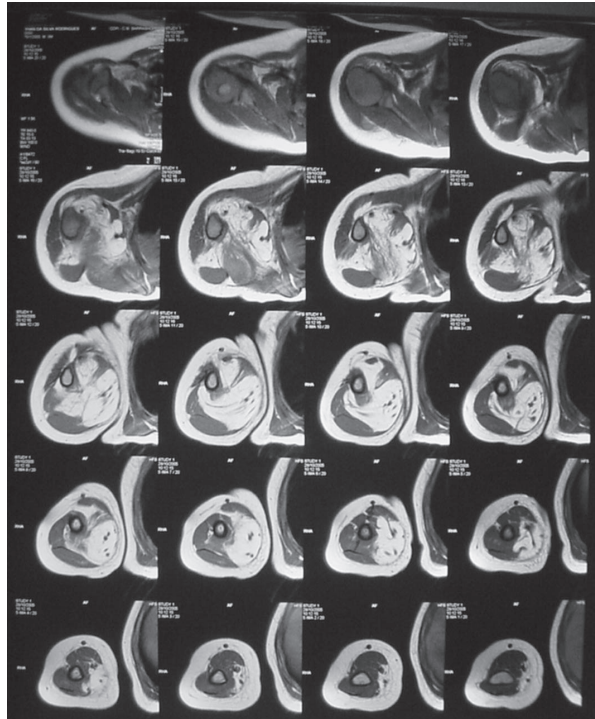


**Figure 1** - Photograph of the lesion at immediate preoperative period.

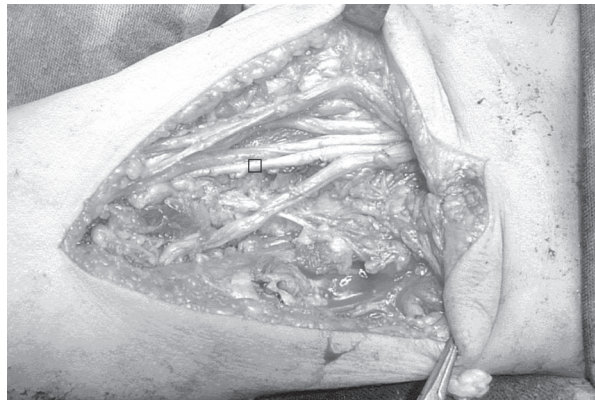
erature. 91% of the cases reported were present during the first year of life, 1/4 of those since birth<sup>(2)</sup>. Male gender is prevalent (2.4:1) and 95% are isolated cases, being commonly present at the axilla, arm, chest, and inguinal/ genital region, but never in hands and feet.

It is painless and grows unpredictably; it may be fast during the early childhood, slowing after the age of 5. There are no reports of spontaneous involution. Malignancy has never been reported, and recurrence after total resection is uncommon (approximately 10% of the cases)<sup>(2)</sup>. Occasionally, trophic changes may occur on supradjacent skin. It is not associated to syndromes or positive family history. Larger lesions typically involve vascular-nervous structures and tumors are infiltrate-like, with undetermined borders and capsule-free, but they do not cross over surrounding structures.

Histologically, these are constituted of mature adipous tissue interposed by portions of a dense fibrous tissue, abundant in myofibroblasts and collagen. They are present next to the portions of connective



**Figure 2** - Cross-section nuclear magnetic resonance, showing the lesion, its relationship with vascular-nervous structures of the arm and humerus.



**Figure 3** - Brachial plexus elements exposed after resection. Marker on brachial artery.

tissue, nests of primitive mesenchyma represented by small immature round cells, with rare mitoses and no atypical areas, sunk in myxoid matrix, possibly constituting an anomalous process of tissue maturation.

The differential diagnosis involves all soft parts tumors, and, considering the hardened tumor fixed to deep planes, it is important to not to confuse it with malign neoplasias, especially to juvenile fibromatosis and sarcomas (particularly rbdomyosarcomas, typical in young children), in order to avoid unnecessary ablative treatments or, correspondently, treatments uniquely resectional or incomplete surgical treatments for malign lesions. Neural tumors (especially neurofibromas) and vascular tumors must also be ruled out<sup>(3)</sup>. In the case of right axillary lesions in our environment, due to the systematic use of BCG vaccine, a differential diagnosis with adenopathies secondary to vaccines is required<sup>(4)</sup>.

Therapeutic approach is the total resection of the lesion, without sacrificing noble structures, considering the low recurrence rate, even in incomplete resections.

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