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Case Report

SOLITARY FIBROUS TUMOR OF THE CHEST WALL IN AN INFANT - A RARE CASE REPORT

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Abstract:

Solitary fibrous tumor is an uncommon mesenchymal neoplasm that very rarely appears in extra serosal soft tissue. Though pleural solitary fibrous tumor is more common, up to one third of the reported cases were at extra pleural sites. Although pleural and extrapleural solitary fibrous tumors are regarded as indolent tumors, there is some evidence that extrapleural subgroup could be a subset of more aggressive malignant tumors. Accurate preoperative diagnosis of extrapleural solitary fibrous tumor is very difficult and usually is a diagnosis of exclusion. Surgical excision is mandatory and is the best therapeutic option. In this article we report a case of solitary fibrous tumor of the chest wall in a 2 months old boy.

Key words : Chest wall tumors, Solitary fibrous tumor, CD34.

Introduction :

Solitary fibrous tumor (SFT) is an uncommon neoplasm.[1,2] Although SFTs mostly occur in a pleural location, up to one-third of cases has been reported in various extrapleural sites such as mediastinum[3], lung[4], meninges[5], prostate, urinary bladder, kidney[6], liver[7], nasal cavity[8], thyroid[9],salivatory glands, upper respiratory tract, peritoneum,spinal cord[10] and esophagus[11]. Extrapleural malignant SFT, especially those arising from chest wall, represent an exceedingly rare subset of soft tissue neoplasms.

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SFT is characterized by a branching, hemangiopericytoma (HPC)-like (staghorn) vasculature. In the past, most SFTs were likely regarded as HPCs.[1,2,12] They typically display zones of both hypercellular and hypocellular collagenized stroma in a so-called patternless architecture.[1,2,12] The cell of origin of SFT has been a matter of contention, and has still not been definitively discerned.[2,12] Although SFTs are usually benign, slow-growing tumors, malignant SFTs have been described, most often in the pleura.[1,2,13-16]

Case presentation

A 2months old boy came to our attention with a protruding mass at the right lateral chest wall at the anterior axillary line overlying the lower ribs. At birth the baby had a slight bulge at the same location and according to the mother one week after birth a small vesicle appeared over the bulge . The vesicle ruptured spontaneously and gradually a red soft tissue mass started to develop over the previous bulge in the chest wall. The baby had no cough or respiratory distress . He was feeding adequately and had significant weight gain. Physical examination showed a slightly tender,firm, red coloured mass with well defined margin overlying the 7th and 8th rib in anterior axillary line. The mass was free from underlying structures and bled on touch. No axillary lymph node was palpable, both lung fields were clear. Chest xray revealed no intra thorasic extension or involvement of the ribs. FNAC from the lump was done and revealed granulation tissue with a special note that possibility of soft tissue sarcoma should be excluded by histopathological examination. Surgical resection was

planned. At surgery the tumour was removed by an elliptical incision (30mm x 30mm x 50mm in size and weighting about 50 grams). No macroscopic evidence of infiltration of surrounding tissue.

Post operative course was uneventful and the patient was discharged on the 3rd post operative day.

Histopathological examination of the excised mass showed a nonencapsulated tumour made of pattern less proliferation of spindled fibroblasts in a keloidal collagenous background with prominent blood vessels resembling hemangiopericytoma but had thin walls ; hyper and hypocellular areas. Mitotic figures were rare to absent. Tumour cells showed cytoplasmic immunoreactivity to CD34.

No further adjuvant treatment such as radiotherapy was given. At present time the patient is alive with no signs of recurrence.

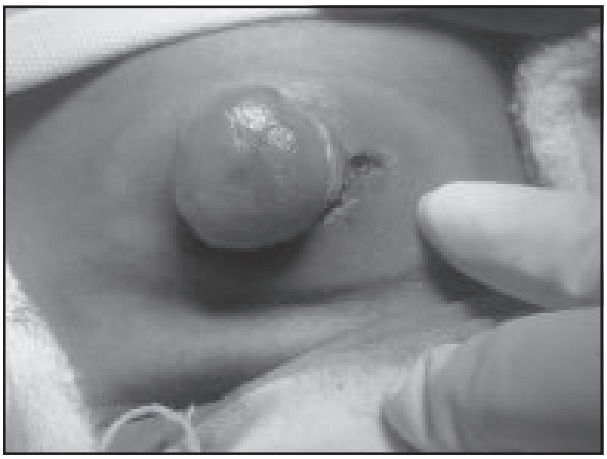

Fig.-1 : *Preoperative appearance of the lump*

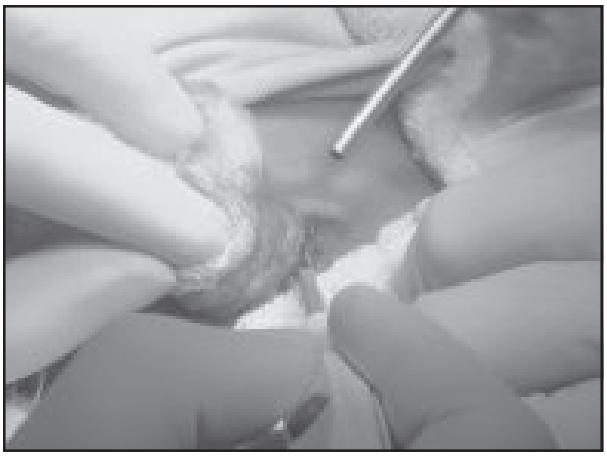

Fig.-2 : *Peroperative dissection*

Fig.-3: *Microscopic features (HE × 200): Round to spindle- shaped cells with little cytoplasm and indistinct cell borders,with vesicular nuclei, sometimes separated by thin bands of collagen.*

Discussion

Solitary Fibrous Tumor (SFT) is an uncommon mesenchymal neoplasm originally described by Klemperer and Rabin with approximately 800 cases reported in the literature between 1931 and 2002[17]. Extrapleural malignant SFT, especially those arising from chest wall, represent an exceedingly rare subset of soft tissue neoplasms. From literature review, only two cases have been described as parascapular lesions[18] and five are described as trunk lesions[19] not well specified as location.

These tumors are observed in middle-aged adults between 25 and 77 years (average age of 45 years) with no sex predilection[20]. They occur only occasionally in children and adolescents. There is no reported incidence of SFT in infants. Sometimes hypoglycemia has been observed in patients with SFTs and this is attributed to the production of insulin-like growth factor II[21], but this represents an ongoing controversial issue. In the present case hypoglycemia was not detected.

Macroscopically SFTs are described as well circumscribed or encapsulated lesions with firm, grey-white cut surface, often pedunculated, and have a remarkable blood supply. Microscopically, they are characterized by an hemangiopericytoma like vasculature "patternless pattern" of growth, and zones of hypercellularity as well as hyopcellularity.[1-2] Typically, strong and diffuse positivity for CD34 and, less significantly for bcl-2 and CD99, is currently regarded as the key finding of SFT.

The majority of these tumors are histologically benign but up to 20% of all SFTs may be malignant. Histological features of extrapleural SFT are essentially similar to those of its pleural counterpart[22].

From literature review, radical surgical resection is mandatory and represents the gold standard. Up to now there is no evidence of effectiveness of chemotherapic and/or radiotherapic adjuvant treatments[23]. Indeed, SFTs have an unpredictable course and, although most of them are characterized by a no aggressive clinical course, locally recurrence or disseminated metastases has been reported[24]. Thus, according with Rovera et al[25] a strict and long term follow-up is recommended.

Conclusion

Chest wall SFTs are a very rare entity with unpredictable behavior. Because of its nonspecific appearance, it is most often not possible to accurately diagnose malignant SFT on FNA material. Precise diagnosis is usually based on the correct interpretation of specific pathologic and immunohistochemical features. Surgical excision as well as a subsequent oncological follow-up are mandatory.

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