A Rare Case of Synovial Sarcoma Involving the Brachial Plexus

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Introduction

Synovial sarcomas are a rare variety of neoplasms accounting for 8% of all soft tissue sarcomas [1]. They are generally found in the paraarticular areas of the tendon sheaths and joints, more commonly the knee joint. However, synovial sarcomas may be found in regions without apparent relationship to synovial structures (<10%) [2, 3].

Head and Neck synovial sarcomas account for less than 0.01% of all malignancies [4].

Only 12 cases of synovial sarcoma involving peripheral nerves have been reported so far.

Three separate histopathological patterns of synovial sarcoma are recognized, viz., monophasic, biphasic and the poorly differentiated type.

Adequate surgical margins, radiation therapy, and adjunct chemotherapy have been reported to correlate with better prognosis [5, 6]. However, despite advances in treatment, mortality rates of synovial sarcoma remains quite high with five-year survival rates around 50–60%.

Case Report

The patient, a 10 year old boy presented with complaints of a swelling in the left supraclavicular region since one year and weakness of left hand since 6 months. The swelling was insidious in onset and was slowly growing initially.

However, it had rapidly increased in size over last six months from a 2 cm diameter to 5.5 cm on the day of admission. On examination, a single, firm, nontender, non-pulsatile supraclavicular swelling of size 5.5 cm was noted posterior to the left Sternocleidomastoid muscle. There was no obvious lymph nodal swelling. Weakness of the Deltoid, Biceps and Brachioradialis and Intrinsic muscles of the hand was noted with reduced sensations in C4, 5, 6, 7, 8, T1 dermatomes. Systemic examination was normal.

MRI of the neck (with contrast) showed the mass to be well defined lobulated, diffusely enhancing with intrathoracic extension, encasing the first rib and the Brachial Plexus as well as the Subclavian vessels with neural foraminal encroachment at C6–C7 and C7–T1 levels, consistent with a benign neurogenic tumour, most likely a neurofibroma (Fig. 1).

FNAC was suggestive of spindle cell tumour. EMG/NCV studies was suggestive of left median, ulnar and radial nerve palsy suggestive of brachial plexus compression.

At surgery, the tumour was found to be arising from C8 root. It was encapsulated, closely adherent to the branches of brachial plexus, extending from under the brachial plexus upto the apical pleura and laterally upto the subclavian artery and medially extending towards the intervertebral foramina of C6–C7 and C7–T1 and seen arising from C8 root. The tumor was moderately vascular and friable. Intracapsular piecemeal excision of the tumour was performed. Almost the entire capsule was removed.

Histopathology revealed it to be a biphasic Synovial Sarcoma, which was confirmed by immunohistochemistry. IHC–EMA, Cytokeratin-7 and 19, S-100, CD-99 were all positive.

The patient underwent Radiation therapy. Over 6 months follow-up, he has complete recovery of power in the upper limb. Coarse sensations had recovered completely.

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Fig. 1 MRI (Neck) shows tumor in close proximity to the Brachial Plexus and Subclavian vessels

Discussion

Synovial sarcomas generally present in young adults around 2nd and 3rd decade of life, with peak incidence at age of 30 years. Our case is rare for the age of presentation of 10 years.

Only 12 cases of synovial sarcoma involving peripheral nerves have been reported till now [7, 8]. Only two cases of synovial sarcoma involving the brachial plexus have been reported so far, one was arising from C7 root and other from T1 root. Our case is 3rd such case, arising from C8 root.

The male: female ratio is around 3:2.

Distant hematogenous metastases are seen with all sarcomas. But unlike other soft tissue sarcomas, lymph nodal metastases is also seen with synovial sarcomas.

Synovial sarcomas are also chemosensitive unlike other soft tissue sarcomas. Hence proper histochemical analysis of these sarcomas is very crucial for proper management of these tumors.

It is difficult to diagnose synovial sarcomas clinically as well as radiologically. Synovial sarcomas are almost always operated upon as being a neurofibroma because they appear to be well encapsulated. In many cases, as the epithelial component is not obtained on aspiration, FNAC can only diagnose that the tumor is a spindle cell tumor. Tru-cut biopsy for histopathological examination and immunohistochemical analysis on the biopsy specimen helps in proper evaluation and treatment planning. Wide excision can be done instead of a piecemeal excision after the proper diagnosis is reached preoperatively.

The most commonly reported overall is the biphasic type, consisting of distinct epithelial cells in gland-like arrangements along with fibroblast-like spindle cell components in varying proportions.

Monophasic tumors consist of spindle cells only and were difficult to distinguish from other soft tissue and nerve

tumors before the introduction of immunostaining for epithelial membrane antigen (EMA), vimentin, S-100, and cytokeratins CK7, CK19, and CD99 [9]. The epithelial components of monophasic synovial sarcomas only become evident when stained for EMA and cytokeratins, although these are not specific for synovial sarcoma. A third histopathological pattern of synovial sarcomas, the poorly differentiated subtype, is also recognized [10]. Although biphasic morphology is most frequently reported overall, eight of the twelve reported intraneural synovial sarcomas have had monophasic histopathology.

Cytogenetic testing is the most definitive way to establish the diagnosis of synovial sarcoma. A stereotyped gene translocation between chromosomes 18 and X t(x;18) (p11.2;q11.2) occurs in over 90% of synovial sarcomas [4]. This leads to fusion of one of two variants of the SSX gene with the SYT gene, resulting in either the SYT/SSX1 or SYT/SSX2 chimeric fusion proteins [9]. Nearly all biphasic tumors express SYT/SSX1, while monophasic tumors express SYT/SSX1 in approximately half of the cases and SYT/SSX2 in the remainder [11]. Patients with SYT/SSX2 expressing tumors have a significantly better prognosis when compared to those with SYT/SSX1 tumors in terms of rates of metastasis and overall survival [12].

While poorly differentiated tumors are associated with a worse prognosis, the prognostic value of monophasic versus biphasic tumors is still debated [13, 14].

Synovial sarcoma is an aggressive tumor that can occur nearly anywhere in the body, including within peripheral nerves as illustrated in our case. Although extremely rare, the potential for a malignancy such as an intraneural synovial sarcoma must be presented in the differential diagnosis to your patient. This is important since the diagnosis may not be apparent until after pathological examination of the surgical specimen, resulting in drastic alterations to the original treatment strategy and an altered impact on the patient.

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