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CASE REPORT

A Rare Case of Synovial Sarcoma Involving the Brachial Plexus, Treated with Wide Local Excision and Reconstructed with Sural Nerve Grafts

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Abstract

This is a case report of monophasic synovial sarcoma involving right brachial plexus. Clinical and radiological examination is not very useful in diagnosis. Histopathology showed spindle cells with occasional mitotic figures. Immunohistochemistry showed positivity for EMA, S-100, Mic-2, Bcl-2, Calponin, TLE-1 and CD-56. Chromosomal translocation t (X;18) (p11.2;q11.2) and chimeric fusion proteins SYT/SSX1 or SYT/SSX2 are formed. Wide local excision of the tumour with involved brachial plexus and later reconstruction of the brachial plexus using sural nerve grafts was done. Patient showed full recovery. Wide excision and radiotherapy is the accepted mode of treatment.

Keywords Synovial sarcoma · Brachial plexus · Head and neck carcinomas · Nerve reconstruction

Introduction

Sarcomas are malignant tumours originating from connective or non-epithelial tissues. Synovial sarcoma (SS) accounts for 8% of soft tissue sarcomas. It is often (not always) found around joints or tendon sheaths. Most commonly involved joint is the knee. Less than 0.01% of all malignancies are head and neck sarcomas. Synovial sarcoma involving peripheral nerves is an extremely rare entity. Only 12 cases have been reported [1].

Case Report

A 27-year-old lady presented with complaints of swelling in right lower neck, paraesthesia of right upper limb and neck pain for 15 days. MRI scan of neck showed a well-defined mass of 9 × 7 mm in lateral interscalene space just posterolateral to anterior scalenus muscle in inferior neck along the anterior division of brachial plexus. Nasopharynx was normal. CT scan of thorax showed no lesions.

Wide local excision of the tumour and anterior divisions of superior and middle trunks, posterior divisions of superior, middle and inferior trunks of right brachial plexus with a margin of 2 cm was done. Repair of the plexus was accomplished with 5 nerve grafts from sural nerve (Fig. 1).

Postoperatively, rest, electric muscle stimulation and later physiotherapy was given till recovery. Function of the involved limb on follow-up after 2.5 years can be seen video.

Discussion

SS are malignant, aggressive and have high recurrence rates. It is more common in males (M:F 3:2) and occurs commonly in the age group of 25–36 years [2]. SS is difficult to diagnose clinically and radiologically. It often presents as a mass which is well circumscribed and covered by thin fibrotic capsule. However, this is a pseudo-capsule beyond which the tumour tends to infiltrate [3]. It is often asymptomatic, and compression on surrounding structures occurs only after tumour gains significant size [4]. It can appear benign and needs to be differentiated from swellings of the neck. CT scans of SS show hypodense, well-demarcated tumours with homogenous

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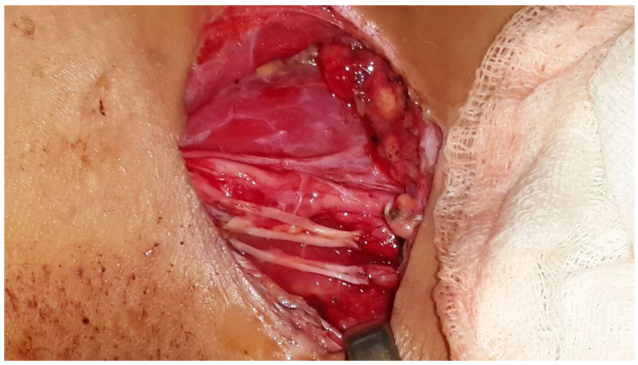

Fig. 1 Postreconstruction photograph of brachial plexus

enhancement. Heterogenous enhancement may be found [5]. On MRI scan of SS of head and neck, T1-weighted images show tumours isointense to grey matter, and T2 weighted images show signal intensity similar to glandular or fat tissue [6]. Histopathology and immunohistochemistry (IHC) helps immensely in diagnosing SS. Three histopathological variants are identified: *biphasic* (most common) consisting of epithelial cells in glandular arrangement with fibroblast-like spindle cell component in varying proportion; *monophasic* shows uniform spindle cell pattern; *poorly differentiated* (least common). Monophasic tumours are commonly < 5 cm while biphasic tumours are > 5 cm in size [7]. IHC of SS are known to be positive for EMA, Bcl-2, CK-7, CD-99 and TLE-1 and negative for CD-34. Bcl-2 is one of the most sensitive markers for SS [8]. Our patient's IHC was positive for EMA, S-100, Mic-2, Bcl-2, calponin (weak), TLE-1 and CD-56 and negative for SMA, Desmin, CD-34, HMB-45 and Melan-A. Thus, diagnosis of monophasic SS was made. Cytogenetic testing can give a definitive diagnosis of SS. In 90% cases, the following translocation occurs t(X;18) (p11.2: q11.2) [9]. This results in fusion of SSX gene with SYT gene giving rise to either SYT/SSX1 or SYT/SSX2 chimeric fusion proteins [8]. SYT/SSX1 fusion is commonly found in the biphasic variant of SS while monophasic variant expresses SYT/SSX1 in approximately half the cases and SYT/SSX2 in the rest. Factors associated with worse prognosis are age > 25 years, tumour size > 5 cm and poor differentiation. Tumours with SYT/SSX2 fusion have a significantly better prognosis than SYT/SSX1 fusion in terms of rates of metastasis and overall survival [10]. The uniformly accepted treatment strategy for SS is wide local excision with negative margins. Postoperative radiotherapy was not required in this case as the primary tumour was < 5 cm. Radiotherapy also delays the recovery of nerve function. So far, only 5 cases of SS involving the brachial plexus have been documented. In our case, wide local excision of the tumour along with involved parts of brachial plexus was done. Reconstruction of the plexus was done using sural nerve grafts. Patient is followed up regularly with improvement of limb function. Role of chemotherapy is reported to be controversial. Overall and 5-year survival rates are 71% and 60% respectively [11]. Our patient was treated in 2015 and is disease free as of 2018.

Conclusion

SS although rare are aggressive tumours which can involve any part of the body including brachial plexus as see in our case. Histopathology and IHC greatly aid in diagnosis of SS which is difficult to diagnose clinically or radiologically. Wide local excision with postoperative radiotherapy forms the mainstay of treatment of SS. The role of chemotherapy still remains debatable.

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