Synovial sarcoma of parapharyngeal and retropharyngeal space: A case report

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Abstract

Synovial sarcoma is a rare tumour of head and neck. It was first described by Pack and Ariel in 1950. Synovial sarcoma is said to be the commonest sarcoma of hands and feet and they are usually found adjacent to the articular surfaces. They rarely occur in extra-articular sites originating from bursa or tendon sheaths. Parapharyngeal space appears to be the site of predilection in most of the cases occurring in the head and neck region. One such case is being reported here along with its management and review of literature.

Keywords Synovial sarcoma · Retropharyngeal and parapharyngeal space lesion

Case Report

A 17-year-old female patient presented at the ENT outdoor, Medical College and Hospitals, Kolkata, with history of gradually progressing swelling of right side of neck for last 9 months, dysphagia for 3 months and hyponasality of voice for last 2 months. There was history of weight loss during last 2–3 months.

Examination of neck revealed a huge swelling of 12.5 X 7.5 cm in size occupying upper part of right anterior triangle of neck extending into the posterior triangle. The swelling was firm, non-tender and multinodular in feel. The skin over the swelling was free and the mobility of the swelling was restricted. The swelling was non-compressible, non-pulsatile and transillumination test was negative. No bruit was heard on auscultation. On examination of the throat, a smooth well-defined swelling was found, occupying right lateral pharyngeal wall pushing the tonsil medially. The swelling also extended to right side of posterior pharyngeal wall. Mucosa over the swelling was normal. The swelling was bimanually palpable. Indirect laryngoscopic examination suggested extension of the swelling to the right pyriform fossa though the lower extent could not be visualised properly. Both vocal cords were mobile. Routine blood and urine investigations were within normal limits. CT scan (Fig. 1) revealed a fairly large oblong soft tissue mass lesion in retropharyngeal plane on the right side of neck deep to the sternocleidomastoid, extending from base of skull above to the C₆ vertebral level below, having calcification in walls in places with bony scalloping in underlying transverse process of C₁.

A neck exploration was performed by a modified Conley's incision. The mass was found adherent to carotid sheath and prevertebral muscle, extending into opposite retropharyngeal space and above up to the base of skull. The whole mass was removed in pieces. The vagus nerve was sacrificed during the procedure. Neck wound was closed in layers. Postoperative period was uneventful except for hoarseness of voice.

The histology of the excised mass showed a highly cellular tumour with biphasic pattern. Gland like spaces lined by epithelial cells were present amidst round to oval spindle shaped cells of small size (Fig. 2). The biphasic pattern was accentuated on reticulin stain. The histological picture suggested the diagnosis of poorly differentiated synovial sarcoma.

The patient was advised postoperative radiotherapy but was lost in follow-up later.

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Fig. 1 CT Scan of the neck of the neck showing heterogenous mass.

Fig. 2 Microphotograph showing the histopathological picture of the excised mass suggestive of synovial sarcoma H & E X 280

Discussion

Synovial sarcoma is a malignant soft tissue tumour thought to arise from a pluripotent mesenchymal stem cell capable of differentiating into cells with epithelial and fibroblastic features. Synovial sarcoma comprise between 8–10% of all malignant neoplasm of somatic soft tissue [1]. Enzinger and Weiss [2] reported 9% incidence of head and neck synovial sarcoma after reviewing 345 cases from Armed Force Institute of Pathology.

In head and neck region, synovial tissues are not plentiful. Synovial tissue in the neck is found in the tendinous portion of the cervical muscle, in the subhyoid

bursa and in the pre-laryngeal bursa. Retropharyngeal and parapharyngeal spaces are devoid of any synovial tissue where most of the sarcoma arises [1]. Most probably, here synovial sarcoma arises from synovial differentiation of primitive mesenchymal cells [3, 4].

The most common symptom of head and neck synovial sarcoma is the presence of a mass. 40% of cases complain of pain. One third of patients in Armed Force Institute of Pathology (AFIP) series presented with complaints of hoarseness, dyspnoea or dysphagia [4]. Synovial sarcoma can occur at any age though it is most commonly observed in adolescents and young adults. 60–70% of cases are found to occur in under 40 age group [2].

The correct pre-operative diagnosis is difficult in cases of synovial sarcoma [1]. CT scan usually shows a heterogeneous pattern with significant contrast enhancement. Calcification is observed in 30% of cases and is considered as a good prognostic sign [5].

Most common pre-operative differential diagnosis is metastatic thyroid carcinoma, which may even mislead the pathologist, who, impressed by the acinar arrangement of the epithelial like components of synovial sarcoma, may consider the lesion as a metastasis from thyroid carcinoma [1]. Other common differential diagnoses at presentation are meningioma, neurilemmoma, lymphoma, chondrosarcoma, nasopharyngeal carcinoma, sarcoidosis, rhabdomyosarcoma in children, mucormycosis in diabetics and aspergillosis in immunocompromised patients [6].

Histologically there are two patterns of synovial sarcoma:

1. Classical biphasic pattern
2. Monophasic pattern (spindle cell variety) which is rare.

The classical biphasic pattern shows two neoplastic components: sarcomatous spindle cell components and gland like clefts lined by epithelial cells. This biphasic pattern is the only histological criterion by which the diagnosis of synovial sarcoma can be made with certainty. The biphasic pattern is enhanced by reticulin stain. The pseudoglandular spaces contain a pink and acidophilic material, which shows positive staining reaction with both PAS and mucicarmine.

In the present case the classical biphasic pattern was present and the initial diagnosis of synovial sarcoma from H&E preparation was confirmed by reticulin and PAS stains. Peripheral and central calcification noted on CT scan was also important corroborative evidence.

Radical surgery is considered the treatment of choice in synovial sarcoma. Radiation therapy has been reported to decrease the high rate of local recurrence. Chemotherapy is useful for control of distant metastasis.

Prognosis of synovial sarcoma in head and neck region relates with size of the tumour [1]. Tumour less than 4 cm in diameter seems to have better prognosis than larger tumours [2]. Improved prognosis is also observed with degree of dif-

ferentiation and calcification [2]. Type of synovial sarcoma usually does not alter the prognosis [1].

Five-year survival rate for head and neck synovial sarcoma is below 50%. In Roth's series survival rate was 47% and most of the patients in Roth's series had pulmonary metastasis at the time of death. Regional lymph node and bony metastasis are common features of peripheral synovial sarcoma though this tendency is not observed in head and neck synovial sarcoma [1].

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