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CASE REPORT

Synovial sarcoma of the mandible

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The term synovial sarcoma was first proposed for the histologically well defined entity by Knox in 1936. The name was proposed due to the fact that at least the cases reported in the early part showed some resemblance of the tumour to normal synovial tissue histologically. Thereafter, tumours have been diagnosed from areas without synovial tissue. The diagnosis of monophasic tumours can be difficult and immunostains appear to be a good diagnostic tool in those cases. The occurrence in the mandible is extremely rare. This is a case of synovial sarcoma arising in the condyle involving most of the ramus. The tumour was diagnosed as synovial sarcoma with predominant spindle cell component by correlation of histopathology with immunostains (cytokeratin, epithelial membrane antigen, actin and desmin). The patient has been treated with surgery and radiotherapy.
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The term synovial sarcoma was first proposed for the histologically well-defined entity by Knox in 1936. The name was proposed due to the fact that at least the cases reported in the early part showed some resemblance of the tumour to normal synovial tissue histologically. However, the origin of the tumour from synovial tissue was not fully substantiated. Although it usually occurs in para-articular regions of extremities, cases have also been reported in relation to para-pharyngeal region, tongue, abdominal wall and buccal cavity where synovial tissue is not present (1).

Approximately 100 cases have been reported in head and neck sites but the occurrence of synovial sarcoma in the mandible is extremely rare. Synovial sarcoma usually occurs in adolescence and young adults between 15 and 40 years of age. This tumour was first described in the head and neck region by Pack in 1950 (2, 3). Because of the rarity of the neoplasm in oro-facial area the possibility of misdiagnosis is a likely event.

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Microscopically, synovial sarcoma shows characteristic biphasic pattern. The usual presentation is that the epithelial cells resembling carcinoma and the spindle cell component with sarcomatous appearance. It is not unusual to see in between forms. This paper documents a case of synovial sarcoma of the right side condyle in a 29-year-old female.

Case report

A 29-year-old female patient presented with a bony hard swelling over the right ramous of the mandible. The swelling has been there for 2 months and gradually increasing in size. The patient was otherwise healthy.

On clinical examination, the swelling was bony hard in consistency with margins merging with the rest of the ramus. There was no evidence of inflammatory signs over the lesion. The patient did not complain of any pain, anaesthesia or paraesthesia in the area. Mouth opening was not restricted.

Radiological features showed a multilocular radiolucency from the neck of the condyle involving most of the right side mandibular ramus. Computed tomography scan confirmed that the tumour appeared to have arisen from right condylar area and it was separated from the Parotid gland.

The tumour was surgically removed and the specimen which measured $6.0 \times 5.0 \times 2.0$ cm was received for histopathological examination. Routine haematoxylin and eosin (H&E) stained sections showed an unencapsulated tumour composed mainly of spindle cells (Fig. 1). In some areas the cells with epithelial features were also observed (Fig. 2). Reticulin stain confirmed the above observation. Cytological atypia was evident in occasional cells. Foci of necrosis were frequently present. The tumour stroma was markedly myxoid. Because of the fact that epithelial component was not marked, the possibility of other spindle cell sarcomas could not be excluded with H&E sections.

Immunostains for cytokeratin (MNF 116, DAKO; Glostrup, Denmark: Fig. 3) and epithelial membrane antigen (DAKO; Fig. 4) were positive in most cells whilst S100 (DAKO) and actin (DAKO) were weakly positive in occasional cells. Desmin (DAKO) was completely negative. The diagnosis of synovial

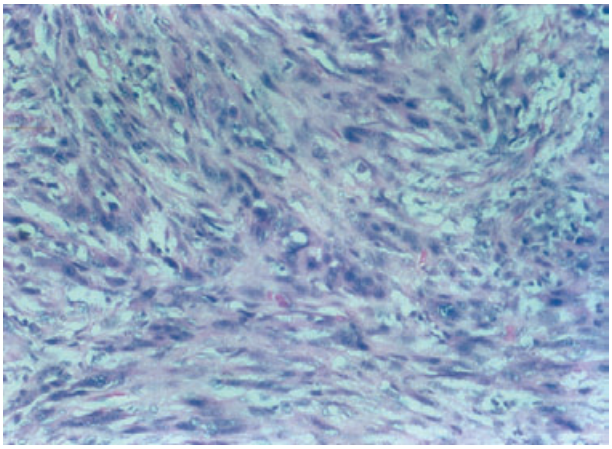

Figure 1 Areas of predominantly spindle cells.

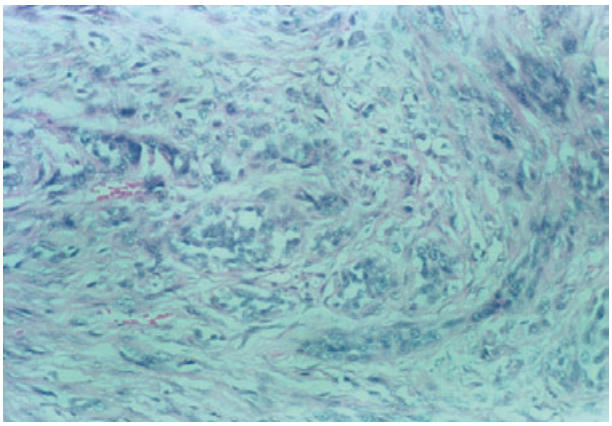

Figure 2 Islands of cells with epithelial appearance.

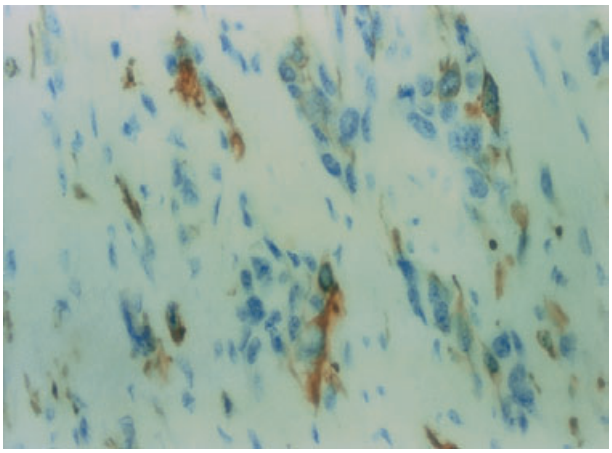

Figure 3 Cytokeratin positive cells.

sarcoma with prominent spindle cell component was arrived with the comparison of H&E features and immunohistochemical findings.

The patient was given adjuvant radiotherapy. The surgical area has healed without complications and the patient is free of recurrence for 2 years of follow-up.

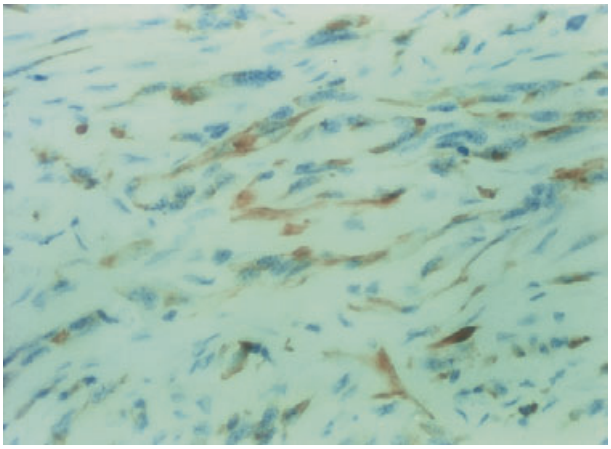

Figure 4 Epithelial membrane antigen positivity.

Comments

This is a case of synovial sarcoma of the mandible in a 29-year-old female who presented with a bony hard swelling of 3 months duration. Histopathological features and immunohistochemical studies were consistent with the diagnosis of synovial sarcoma.

Although synovial carcinoma primarily occurs in para-articular sites of extremities, head and neck may be a site for some cases. The commonest site of occurrence of the tumour in the head and neck region is the pharyngeal area. A very few cases have been reported inside the oral cavity. The majority of synovial sarcomas in the head and neck occur in young adults but sporadic cases have been reported in children. Synovial sarcoma contains characteristic chromosomal translocation, t(x; 18) p11.2j q11.2, which is a useful diagnostic tool especially when histological features are equivocal (3).

Radiological features of synovial sarcoma are some times helpful to arrive at a reasonable differential diagnosis. They show round to oval radiolucency without major bone destruction. The underlying bone tends to be uninvolved but 15–20% will show periosteal bone reaction. Focal calcification is a feature in 15–20% of the cases. The tumour in the head and neck region may not show these characteristic features. Rarity of the lesion and atypical radiological features in head and neck sites may not allow the clinician to arrive at a radiological diagnosis of synovial sarcoma.

Diagnosis of typical synovial sarcoma may not be a challenging task, especially when the biphasic pattern of the cells are present. However, synovial sarcoma has a broad spectrum of histological features. In addition to the classic biphasic type, monophasic, epithelioid monophasic fibrous and poorly differentiated types have been described. Histological differential diagnosis varies with the sub-type of synovial sarcoma.

Monophasic tumours can resemble fibrosarcoma, leiomyosarcoma, malignant peripheral nerve sheath tumour, haemangiopericytoma, and some times malignant fibrous histocytoma. Especially in the head and

neck synovial sarcoma with round cells may be confused with any of the small round cell tumours (4). Therefore it is advisable to use a panel of antibodies and immunohistochemical findings must be assessed together with other observations. Cytokeratin positivity is found in about 75% of the tumours and epithelial membrane antigen also shows the same sensitivity as cytokeratin (5). Expression of cytokeratin is usually evident in epithelial cells. However in some cases it has been reported that the non-epithelial cells also stain for cytokeratin (5). The present case also showed that occasional spindle cells to be positive for the pan keratin marker (MNF116). Synovial sarcoma shares keratin 8 and 18 with other spindle cell sarcomas, but keratin 7, 19 and desmoplakin seem to be more specific to the epithelial component of the tumour (7).

Usually local recurrence is expected within 2 years of diagnosis but can appear up to 20 years. Metastases are usually blood borne to lungs (94%). Twenty-five percent of the cases can involve regional lymph nodes.

According to the current understanding, tumours of the limb/limb girdle, young age, low mitoses ($< 15/10$ HPF) and presence of calcification are regarded as factors which favour better prognosis.

Five year survival rate is 25–50% and the 10 year survival rate is only 10–15%. Five year survival rates are however higher with the modern therapy including adequate excision, irradiation, adjuvant chemotherapy and surgical metastasectomy. Accordingly some authors have shown 5 year survival rates of 65–70% (6). Sites, which are less accessible such as head and neck, are less amenable to cure.

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