MALIGNANT SCHWANNOMA OF MANDIBLE - CASE REPORT

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SUMMARY: A case of malignant schwannoma of the mandible in a forty year old female is presented because of its rarity at that site. The clinical features, histopathology and management is presented and the relevant literature is reviewed.

INTRODUCTION

Malignant schwannoma is rare in the maxillo-facial region. Usually it occurs in association with Von Recklinghausen’s disease of nerves. Gratz, in 1991, has mentioned that only 26 cases of malignant schwannoma of the oral cavity have been reported. We are reporting a case of malignant schwannoma of the mandible not associated with Von Recklinghausen’s disease.

CASE REPORT

A forty year old lady presented with painless swelling of the right side of the lower jaw which had been present for eight months. There was no history of paraesthesia, previous trauma or toothache. She had undergone extraction of an impacted right lower third molar ten years back.

On examination there was a fusiform swelling of the mandible which extended from the subcondylar region to the first premolar on the right side, expanding both tables of the mandible. Skin and mucosa over the swelling were not involved. The swelling was firm and non tender. Regional lymph nodes were not enlarged and there was no facial paralysis (Fig. 1).

Skingram of the mandible revealed a cystic lesion extending from the condylar region to the first premolar. Few trabeculas were seen in the cyst. There was no bony sclerosis or areas of osteolysis. Mandibular canal was not clearly visualised (Fig. 2).

Clinical diagnosis of adamantinoma was made and hemimandibulectomy was planned. By a submandibular incision the part of the mandible including the swelling from the condyle up to the right canine was resected. The defect in the mandible was primarily reconstructed by iliac bone graft. The resected specimen measured 11 cm x 4 cm.

Histopathology revealed a tumour showing spindle cells arranged in interlacing bundles with elongated nuclei. Stroma showed wavy neural fibres. There were focal areas of hypercellularity, hyperchromasia and increased mitotic activity with mild pleomorphism. A few normal bony spicules were infiltrated by tumour cells suggestive of malignant schwannoma (Fig. 3 & 4).

Based on the histopathology findings, radiotherapy was started to the area from the tenth post
operative day, 5700 rads were given in fractionated doses over a period of forty days. On one and half years follow up she is recurrence free (Fig. 5 & 6). Orthopantomogram reveals a well consolidated iliac bone graft (Fig. 7).

**DISCUSSION**

The term "malignant schwannoma" is used to designate malignant tumours of peripheral nerve sheath origin. Most people feel that this tumour originates from Schwann cells; but some studies point out to a possible origin from perineurial cells or fibroblasts. Some peripheral nerve sheath tumours have histological features of fibro sarcoma and hence have been termed as neurofibrosarcoma. These tumours constitute 2-12% of all nerve sheath tumours. They are more common in males and occur at an early age of thirty years when associated with Von Recklinghausen's disease. When a spontaneous malignant tumour arises, it is more common in females and occurs in the age group of 40 to 45 years. They usually present as a painless expanding mass. They usually arise in the trunk, extremities and in the paravertebral region. Gratz, in a review, has mentioned that only 26 cases of malignant schwannoma have been reported in oral cavity. Only a few are reported in the mandible. Radiologically, malignant schwannoma can be classified into two varieties. There can be a cystic lesion in the mandible or an
obvious cylindrical enlargement of mandibular canal. Electron microscopy shows well differentiated spindle cells with intervening cytoplasmic processes, micro filaments and micro tubules. Immunohistochemical studies demonstrate S-100 protein in some cases and vimentin in 90% of the cases. Type IV collagen is present.

Malignant schwannomas are highly aggressive tumours with a tendency to recur locally in a short time. They may even recur 5 to 10 years after primary treatment. Recurrence rate after local excision is 46%. Prognosis is poor when the tumour occurs in patients with Von Recklinghausen's disease and when the tumour arises in inferior alveolar nerve.

References

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