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Pathology in Focus

Primary osteogenic sarcoma of the tongue

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Abstract
A 56-year-old man presented with the difficulty of swallowing and respiratory distress due to a large tumour arising from the tongue and occupying the entire oral cavity. Histological examination revealed it to be an extraskeletal osteogenic sarcoma. The tumour was excised. After six weeks, he came back with massive local recurrence and bleeding from the tumour, but died despite chemotherapy. Review of the literature revealed only four other such cases of this rare tumour. A brief review of these four cases is also made.

Key words: Tongue neoplasms; Osteosarcoma

Introduction

Osteogenic sarcoma of the extraskeletal soft tissues is a rare pathologic process. It has been reported to arise in many parts of the body (Allan and Soule, 1971; Sordillo *et al.*, 1983; Chung and Enzinger, 1987). We encountered such a tumour arising in the tongue of a 56-year-old Melanesian patient. The present report describes our experience of the clinical features, pathologic nature, treatment and prognosis of this rare neoplasm. A review of the very few such cases available in literature is also made.

Case report

A 56-year-old male patient, was referred to the Otolaryngology clinic of the Port Moresby General Hospital with complaints of a gradually increasing swelling on the tongue with difficulty in swallowing for seven months, and increasing respiratory distress for two months. He had a past history of moderate alcohol consumption, tobacco smoking and raw betel nut chewing. Clinical examination showed a tumour, with a nodular surface, firm to hard in consistency, with areas of superficial ulcerations, arising from the dorsum of the tongue and occupying the entire oral cavity. The tongue was mobile. Fibre-optic laryngoscope showed, (i) secretions in both nasal cavities, (ii) upward bulging of the soft palate and uvula, and (iii) the tumour mass sitting over the base of the tongue, the valecullae, and abutting against the lingual surface of the epiglottis and the posterior pharyngeal wall. These findings were more pronounced on the right side. Further down the larynx appeared normal. On physical examination, he appeared to be of his stated age and in marked respiratory distress. There was no regional lymphadenopathy and the rest of the physical examination, including the skeletal system, were within the normal limits. Haematological tests, including alkaline phosphatase, were normal. No other primary or secondary neoplasm was found anywhere in the body. X-ray of the lateral view of the neck showed soft tissue opacity in the oral cavity and oropharynx. The latter extended posteriorly up to the posterior pharyngeal wall and inferiorly up to the epiglottis. The admission chest radiograph was normal. The radiograph of the mandible revealed no bony involvement by the tumour. The skeletal survey of the body did not reveal any abnormality. A biopsy of tumour mass was carried out at the time of the emergency tracheostomy. At a later stage right hemiglossectomy by right paramedian mandibulotomy was per-

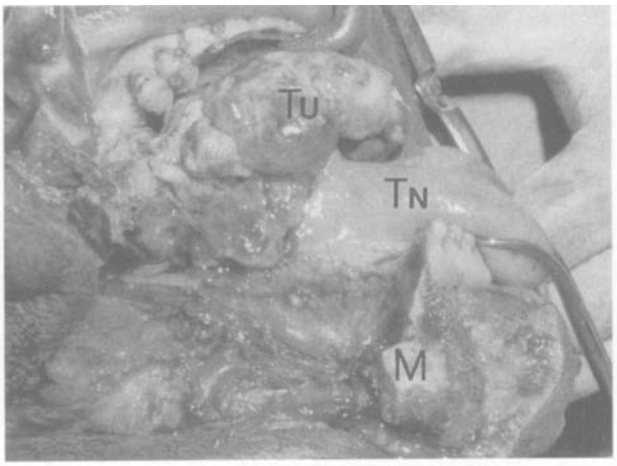

FIG. 1
Intra-operative photograph after right paramedian mandibulotomy showing the tumour (TU) on the dorsum of the tongue (TN); M is the cut larger segment of the mandible.

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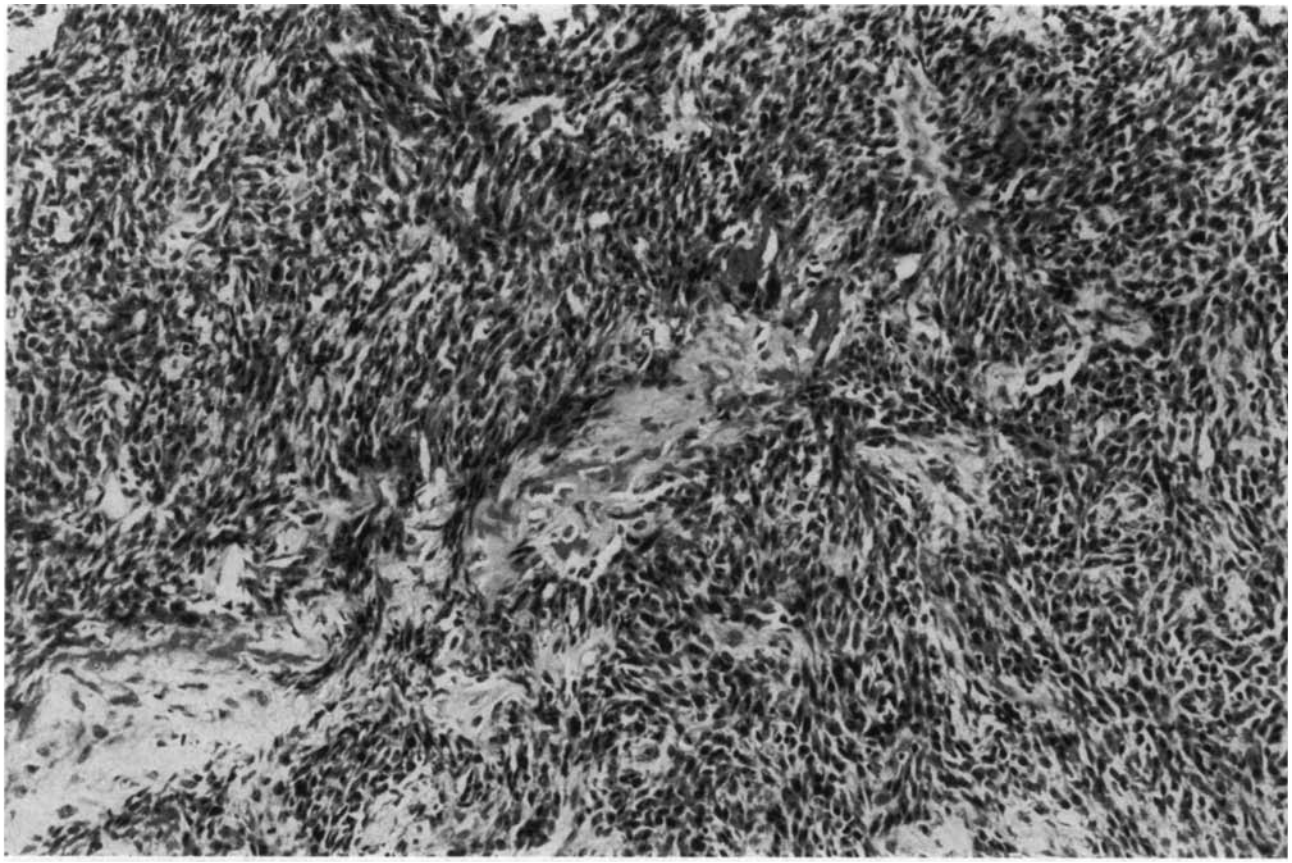

FIG. 2

Section of the tumour with a spindle-cell pattern with hyper-chromatic nuclei and foci of osteoid formation. (H & E; × 100)

formed (Figure 1). The tumour was found to be attached to the right half of the dorsum of the tongue from the mid-part of its anterior two thirds, the right lateral margin and right half of the posterior one third. The tumour was friable and was fragmented during its removal. The resultant defect was repaired and the wound was closed after wiring the step mandibulotomy. During the surgical procedure no mandibular involvement by the tumour was noted. There was no additional attachment of the tumour to tissues other than those described earlier. The patient's subsequent recovery was uneventful. The patient refused post-operative irradiation.

Six weeks following operation, the patient developed massive local recurrence of the tumour and intermittent moderate bleeding from it. Repeat skeletal survey and serum alkaline phosphatase were within the normal limits. The patient was treated with one cycle of chemotherapy consisting of cyclophosphamide, methotrexate and adriamycin. But there was no reduction of the size of the tumour. The patient's condition deteriorated very fast and he died. Permission for autopsy was not granted.

The initial incisional biopsy of the tumour at the time of emergency tracheostomy consisting of small soft white tissue fragments averaging about 5 × 5 × 3 mm size revealed a spindle-cell malignant tumour with foci of hyalinization and was interpreted as soft tissue sarcoma.

The hemiglossectomy specimen consisted of four irregular nodular yellowish-white to brown firm tissue pieces of 8 × 6.5 × 4 cm, 6 × 4 × 4 cm, 5.5 × 3.5 × 3 cm and 4 × 2.3 × 2 cm and five smaller tissue pieces of similar nature ranging from 1 × 0.5 × 0.5 to 2 × 1.5 × 1 cm size. The cut surfaces of the tissue pieces were white and nodular.

Multiple sections of the formalin-fixed paraffin-embedded tissue blocks stained with haematoxylin and eosin showed ulceration on the surface of the lesion, non-specific inflammation in the superficial region and a tumour underneath. The tumour was composed of a spindle and polygonal cells with large nuclei with prominant nucleoli (Figure 2). Mitotic activity, abnormal mitoses, tumour giant cells and cellular pleomorphism were observed (Figure 3). Foci of osteoid and bone formation were present in the tumour in many areas (Figure 4). The tumour exhibited a uniform pleomorphic morphology in all the sections. The tumour was diffusely infiltrating into the muscle and the adjacent tissues. The features were consistent with osteogenic sarcoma.

Immunoperoxidase stains for keratin, CAM 5.2 and S100 were negative. The stain for vimentin was very strongly positive, with virtually all of the tumour cells staining positively. The negative staining for keratin, CAM 5.2 and S100 rule out the possibility of this tumour being of epithelial or neural origin. The strong reaction for vimentin indicates the mesenchymal origin of the tumour. In the absence of any demonstrable connection with the bone, we consider this tumour as a primary soft tissue osteosarcoma arising in the tongue.

Discussion

Extraskeletal osteogenic sarcoma is a well recognized but rare soft tissue neoplasm and in a series reported from the Mayo Clinic this tumour made up 1.2 per cent of all soft tissue sarcomas (Allan and Soule, 1971). Whereas osteosarcomas of bone occurs most frequently in children and adolescents, patients with extraosseous osteogenic sarcomas are almost always over 40 years of age, usually in the fifth or sixth decades of life (Allan and Soule, 1971; Sordillo *et al.*, 1983; Chung and Enzinger, 1987). The rest of its biological behaviour is similar to those of skeletal osteogenic sarcomas (Sordillo *et al.*, 1983). Despite

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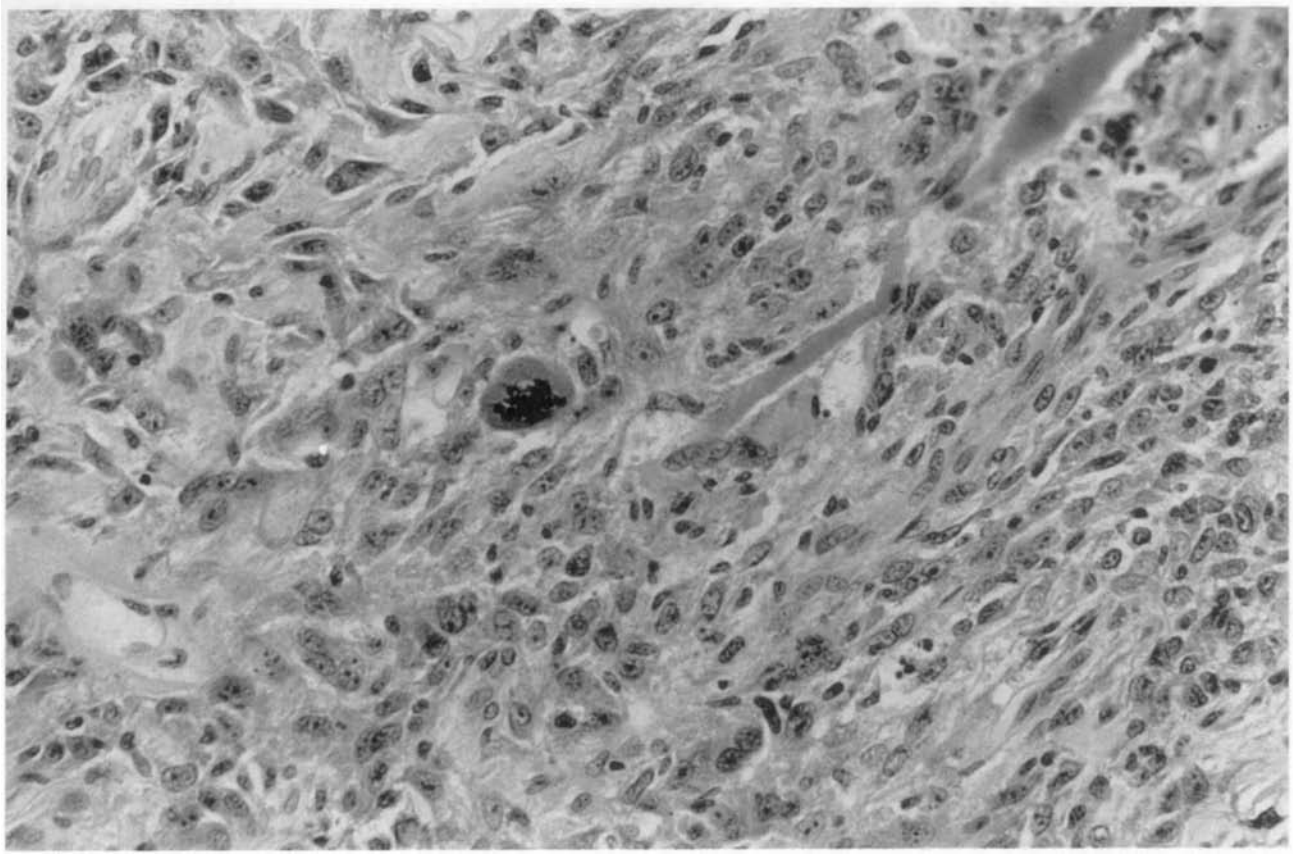

FIG. 3
Section of the tumour with tumour giant cell and nuclei with nucleoli. (H & E: × 400)

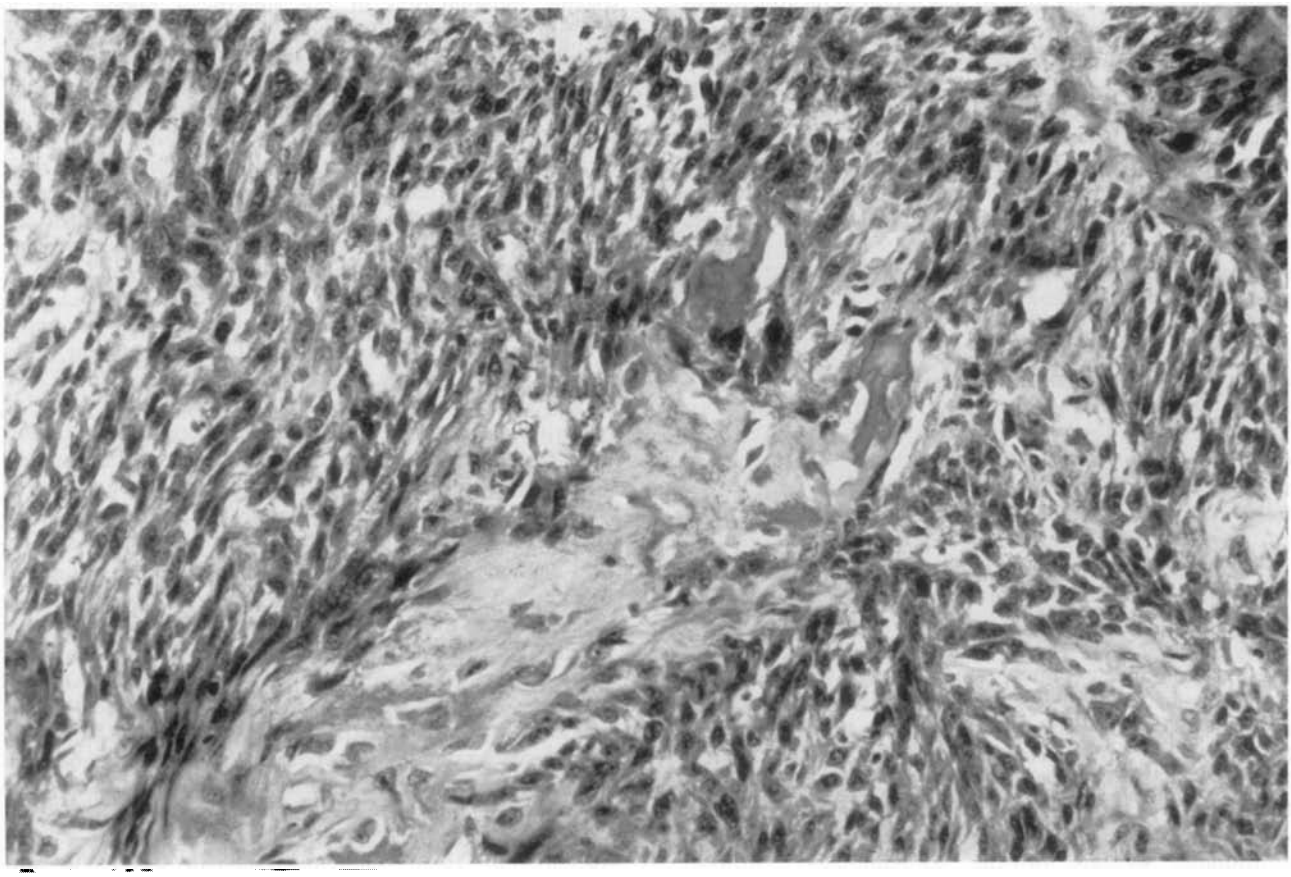

FIG. 4
Section of the tumour with a large area of osteoid and a small bony spicule within the focus of osteoid. (H & E: × 200)

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TABLE I
CLINICAL DETAILS, TREATMENT AND FOLLOW-UP DATA OF PREVIOUSLY REPORTED CASES

Authors/ Year	Sex/age (yrs)	Presentation	Location	Size	Additional information	Treatment	Follow-up
Reyes *et al.* (1981)	F/45	Pain, difficulty in swallowing for 2 weeks	Left lateral margin in the middle third	7 × 5 × 4 cm	Bilateral pulmonary metastases	Hemiglossectomy, chemotherapy (cytoxan, methotrexate, adriamycin)	Died after 27 days of hospitalization
Bem and Sharpe (1988)	M/64	Swelling and difficulty in protruding the tongue for 3 months	Mobile, ulcerated mass in the posterior third	—	Combined interstitial (59.10 Gy) and external (62.70 Gy) irradiation for carcinoma tongue 25 years ago	Total glossectomy	No recurrence up to 9 months post-operatively
Beziat *et al.* (1989)	M/56	Pain, otalgia, difficulty in swallowing for 3 weeks	Left posterior part of the border of the tongue	3 cm in diameter	Alcohol consumption, tobacco smoking	Hemiglossectomy, triangular neck dissection, 60 Gy irradiation, chemotherapy (adriamycin, cyclophosphamide)	No recurrence up to 4 years post-operatively
Loyzaga *et al.* (1996)	M/81	Ulcerated lump of the tongue, palpable neck node	Posterior part of the left border	4.5 × 4 × 3.5 cm	—	Hemiglossectomy, neck dissection	No recurrence up to 25 months post-operatively

combination therapy with surgery, irradiation and chemotherapy, the long-term survival of these patients is very poor in the most reported series (Allan and Soule, 1971; Sordillo *et al.*, 1983; Chung and Enzinger, 1987). The usual course of this disease in most patients is that of multiple local recurrences, followed by distant metastases and death (Allan and Soule, 1971; Sordillo *et al.*, 1983; Chung and Enzinger, 1987).

If extraskeletal osteosarcoma, in general, is considered as a rare tumour, its location in the tongue is exceptional. A 36 years review from 1946 to 1982 of 88 cases in the Armed Forces Institute of Pathology, Washington, revealed only 4.5 per cent incidence of this neoplasm in the head and neck (Chung and Enzinger, 1987). Because of its rare occurrence in the tongue, little information is available regarding its relative incidence, clinical behaviour and result of treatment in contemporary literature. A review of the world literature revealed only four other cases of primary osteogenic sarcoma of the tongue (Reyes *et al.*, 1981; Bem and Sharpe, 1983; Beziat *et al.*, 1989; Loyzaga *et al.*, 1996). The clinical details, treatment modalities and the follow-up data of these four cases are given in the table (Table I).

Histological differential diagnosis of extraskeletal osteosarcoma includes malignant fibrous histiocytoma and myositis ossificans (Chung and Enzinger, 1987). The features of osteoid and bone formation within the malignant spindle-cell sarcomatous tumour with uniform morphology demonstrated in our case excludes malignant fibrous histiocytoma and meet the diagnostic histological criteria suggested by Stout and Lattes for extraskeletal osteosarcoma (Stout and Lattes, 1967). The absence of connection of the tumour with the mandible also fulfils further the criteria described by Stout and Lattes (1967). The features of osteiod and bone formation were observed throughout the lesion. Absence of zonal arrangement, trabecular and marginal deposits of osteoid and metaplastic demarcation of bony and chondroid nests in the tumour exclude the diagnosis of myositis ossificans in our case. This is further supported by the absence of history of trauma.

Conclusion

Extraosseous osteogenic sarcoma is a rare malignant neoplasm that can occur in any tissue or organ in the body. The tongue is quite an exceptional site. All such cases must be recorded in the literature. This will help to gain experience about this tumour with respect to its clinico-pathological features, diagnosis, appropriate treatment modalities and survival.

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