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Extramedullary plasmacytoma

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EXTRAMEDULLARY plasmacytomas form a small but important fraction of malignancies in the head and neck region. Although these tumours are rare, the head and neck are the commonest regions involved. The literature over the years has contained case reports from various centres and all have emphasized the rarity of these tumours (Webb *et al.*, 1962; Helmus, 1964; Booth *et al.*, 1973; Batsakis, 1974; Pahor, 1977).

This report deals with three cases seen during a period of three years; one each in the nose, larynx and maxilla.

Case reports

Case 1. A 50-year-old farmer was admitted in July, 1975 with complaints of nasal obstruction and swelling over the nose for 6 months. There was no history of epistaxis or rhinorrhoea. Clinical examination showed a large rounded swelling over the bridge of the nose, 8 cms. × 8 cms., in size, involving the inner canthus of both eyes. The mass was firm and non-tender with the skin stretched over it (Figs. 1 and 2). Anterior rhinoscopy showed a greyish pink mass filling both nostrils. Posterior rhinoscopy revealed a similar mass coming out of both choanae. The oral cavity was normal. There were no palpable cervical lymph nodes and the patient appeared to be in good general health.

Routine blood and urine examination and paranasal sinus X-rays were within normal limits. Biopsy from the nasal mass was reported as Plasmacytoma. Further investigations were conducted to rule out multiple myeloma. Skeletal survey was unrewarding. There were no Bence-Jones proteins in the urine, and the plasma proteins and bone marrow studies were also normal.

The growth was treated with megavoltage radiotherapy. A dose of 5,500 rads. was given over 5 weeks, and this resulted in a marked improvement with no residue at the time of discharge in November 1975. The patient is being followed up, free of disease.

Case 2. A 42-year-old male came to us in July, 1976 with a complaint of hoarseness of voice of one year's duration. There was no cough, dyspnoea or dysphagia. The hoarseness started suddenly and was progressive. The patient was treated elsewhere, but without any improvement.

Examination by indirect laryngoscopy revealed a large pinkish smooth mass in the right ventricle, causing a bulge in the false cord on the same side. The right true cord could not be seen because of the mass. Blood and urine examinations were normal.

After a preliminary tracheostomy, a laryngofissure was done to remove the mass from the right ventricle, and the pathologists reported it to be a plasmacytoma (Fig. 3). Further investigations to rule out multiple myeloma, such as a skeletal survey, urine examination for Bence-Jones proteins, serum protein examination and sternal marrow biopsy were unrewarding.

The patient improved, but a laryngogram done 3 weeks after the excision

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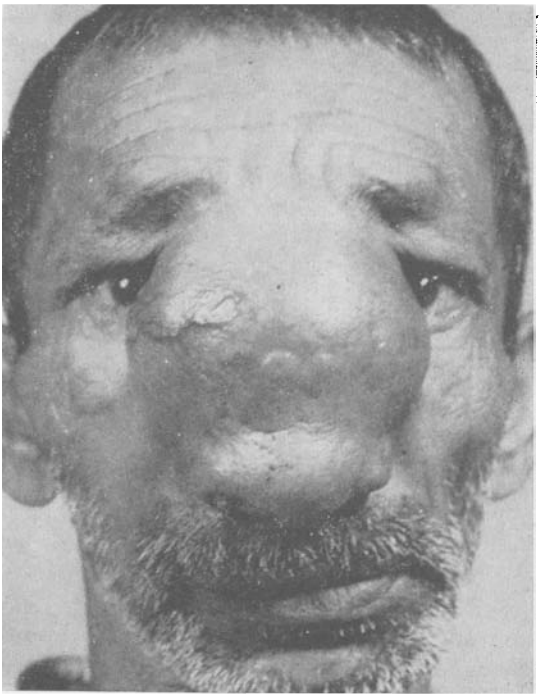


FIG. 1.
Front view of patient 1.

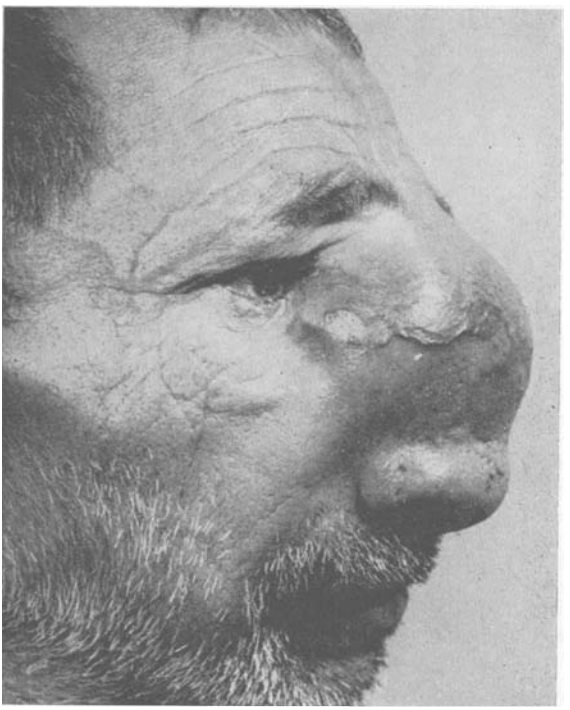


FIG. 2.
Right-sided view of patient 1.

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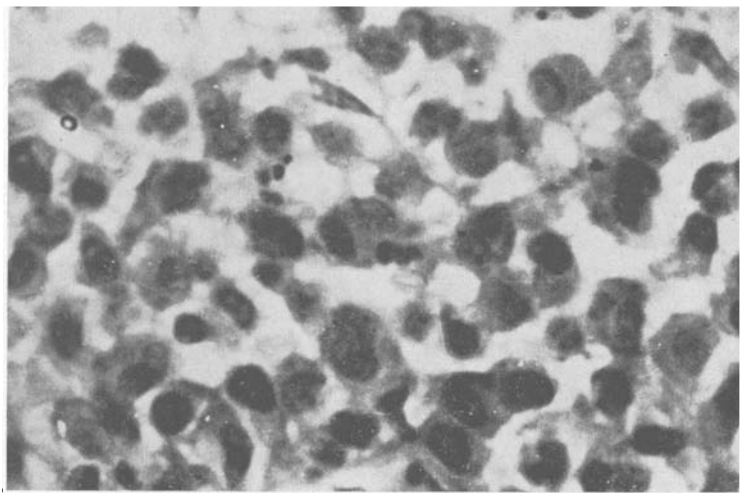


FIG. 3.

Histopathological photograph of case 2. (Haematoxylin-Eosin stain, magnification ×200).

showed restricted opening of the right ventricle. The patient was given 5,000 rads. of megavoltage radiotherapy in 5 weeks, after which the right ventricle showed normal outlines. The patient was perfectly well when last seen in December 1978.

Case 3. A 60-year-old female was admitted in November, 1976 with a swelling over the left hard palate of 2 months' duration and a swelling over the left side of the face for 2 weeks. The swelling on the palate was preceded by loosening and falling out of upper teeth on the same side. There was no complaint of nasal blockage or epistaxis. Examination showed a diffuse hard swelling over the antero-lateral surface of the left maxilla, without any abnormality of the infraorbital margin. On examining the oral cavity, a firm cystic bulge, 5 cms. × 5 cms., was present on the left side of the hard palate and alveolar process, with absence of teeth in the area (Fig. 4). The biopsy was reported as Plasmacytoma. The presence of multiple myeloma was ruled out by further investigations.

A left total maxillectomy was done in February, 1977. Her recovery was uneventful and she was kept under constant follow-up. In March 1978, the patient developed a swelling over the left zygomatic region, 4 cms. × 4 cms., hard and fixed to the underlying bone, but the overlying skin was free (Fig. 5) and the maxillectomy cavity was healthy. The patient was treated with 6,000 rads. in 6 weeks, and discharged. The patient remained disease-free at a recent follow-up.

Discussion

Solitary extramedullary plasmacytoma is a distinctly uncommon tumour. These lesions occur chiefly in the upper respiratory and alimentary tracts, including the nares, sinuses, tonsils, palate, tongue, gingiva and floor of the mouth. They have also been found in the pleura, mediastinum, thyroid, spermatic cord, ovary, intestines, kidney and skin (Lucas, 1972). The head-and-neck region, however, is the one most commonly involved. Figures of 75 per cent

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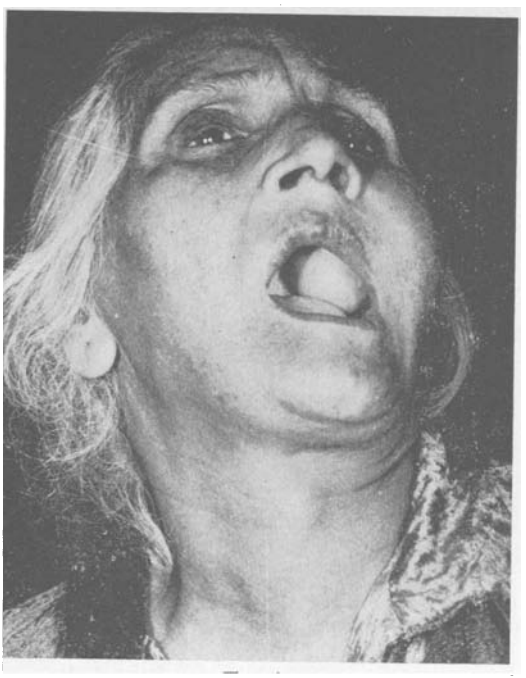


FIG. 4.

Photograph of case 3, showing the swelling over the left cheek and palate.



FIG. 5.

Photograph of case 3, showing recurrence over the left zygoma.

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(Wiltshaw, 1971), 80 per cent (Batsakis, 1974) and 90 per cent (Booth *et al.*, 1973) have been given regarding the involvement of the head and neck region.

Generally the diagnosis of plasmacytoma comes as a surprise following biopsy, since there is nothing distinctive about the gross appearance of the tumour. Once such a report is made, further investigations must be carried out to rule out the presence of a generalized form of multiple myeloma. Absence of other radiologically proved lesions in the skeletal survey, negative sternal marrow biopsy and absence of Bence-Jones proteinuria are the criteria for the diagnosis of such a lesion. Azar and Potter (1973) believe that the validity of a diagnosis of solitary myeloma is often disputed on the grounds that, in the absence of an autopsy or of a long follow-up, no case of solitary myeloma can be definitely proved to be so. The extramedullary manifestations may long precede a clinically occult but nevertheless disseminated myelomatosis. Willis (1941) and Raven and Willis (1949) believe that a lesion can be accepted as solitary only if radiographic examination of the skeleton fails to show other deposits for at least one year after the original diagnosis. Only one of our cases showed a local recurrence, but none developed a generalized dissemination of the disease.

In the head-and-neck region there is no portion that is immune, but there is a predilection for the nasopharynx, nasal cavity, paranasal sinuses and tonsils. Many authors (Hellwig, 1943; Stout and Kenney, 1949; Dolin and Dewar, 1956) believe that the nose is the commonest site, while Webb *et al.* (1962) believe the maxillary sinus to be a more common site.

Extramedullary plasmacytoma involving the larynx is a still rarer entity. Helmus (1964) reviewed the literature and found 150 cases of plasmacytoma of which only 9 were in the larynx. Webb *et al.* (1962) reported 19 cases of plasmacytoma from the Mayo Clinic and only 3 of these were cases of laryngeal tumours. Poole and Marchetta (1968) reviewed 104 cases of upper respiratory tract plasmacytoma and found an incidence of 4·5 per cent in the larynx. Cady *et al.* (1968) reviewed 2,500 cases of cancer of the larynx from the Memorial Hospital, from 1940–1965, and found only one case of plasmacytoma. More cases have been reported by Booth *et al.* (1973), Griffiths and Brown (1974) and Gorenstein *et al.* (1977). In our case, the patient was found to have a lesion in the ventricle.

The recommended therapy is uncertain. Its rarity makes it difficult for any one medical centre to accumulate a sufficient number of cases to be statistically meaningful (Dodson, 1975). Helmus (1964) reviewed the results of treatment by surgical excision, irradiation therapy and a combination of the two; and he concluded that no statistically significant difference could be demonstrated. The method of treatment is guided by the site of involvement and by the response. Sisson and Goldstein (1973) favour local resection where possible, with extensive electro-coagulation of the tumour bed and margins; and they prefer to reserve radiotherapy for recurrences.

Of the presented series, the case with the nasal mass (case 1) was treated with radiotherapy and was completely cured. The case with the tumour in the maxilla (case 3) was treated by total maxillectomy, but the patient developed a zygomatic lesion which was treated with radiotherapy. The one with the laryngeal involvement (case 2) was treated initially by surgical excision, but a doubtful residue was treated with a short course of radiotherapy.

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Summary

Extramedullary plasmacytomas of the head and neck are very uncommon tumours. Three rare cases of extramedullary plasmacytoma are presented—one each in the nose, larynx and maxilla.

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REFERENCES

AZAR, H. A., and POTTER, M. (1973) *Multiple Myeloma and Related Disorders*. Maryland, Harper and Row. Vol. 1, p. 16.

BATSAKIS, J. G. (1974) *Tumours of the Head and Neck*. Baltimore, Williams and Wilkins, p. 362.

BOOTH, J. B., CHEESMAN, A. D., and VINCENTI, N. H. (1973) Extramedullary plasmacytoma of the upper respiratory tract. *Annals of Otology, Rhinology and Laryngology*, **82,** 709.

CADY, B., RIPPEY, J. H., and FRAZELL, E. L. (1968) Non-epidermoid cancer of the larynx. *Annals of Surgery*, **167,** 116.

DODSON, T. A. (1975) Solitary extramedullary plasmacytoma of the head and neck. *Transactions American Academy of Ophthalmology and Otolaryngology*, **80,** 472.

DOLIN, S., and DEWAR, J. P. (1956) Extramedullary plasmacytoma. *American Journal of Pathology*, **32,** 83.

GORENSTEIN, A., NEEL, H. B., DEVINE, K. D., and WEILAND, L. H. (1977) Solitary extramedullary plasmacytoma of the larynx. *Archives of Otolaryngology*, **103,** 159.

GRIFFITHS, C., and BROWN, G. (1974) Extramedullary plasmacytoma. *Canadian Journal of Otolaryngology*, **3,** 81.

HELLWIG, C. A. (1943) Extramedullary plasma cell tumours as observed in various locations. *Archives of Pathology*, **36,** 95.

HELMUS, C. (1964) Extramedullary plasmacytoma of the head and neck. *Laryngoscope*, **74,** 553.

LUCAS, R. B. (1972) *Pathology of Tumours of the Oral Tissues*. London, Churchill Livingstone. 2nd Ed. p. 233.

PAHOR, A. L. (1977) Extramedullary plasmacytoma of the head and neck, parotid and submandibular salivary glands. *Journal of Laryngology and Otology*, **91,** 241.

POOLE, A. G., and MARCHETTA, F. C. (1968) Extramedullary plasmacytoma of the head and neck. *Cancer*, **22,** 14.

RAVEN, R. W., and WILLIS, R. A. (1949) Solitary plasmacytoma of the spine. *Journal of Bone and Joint Surgery (British)*, **31,** 369.

SISSON, G. A., and GOLDSTEIN, J. C. (1973) Tumours of the nose, paranasal sinuses and nasopharynx. In *Otolaryngology*. Editors Paparella M. M. and Shumrick, D. A. Philadelphia, W. B. Saunders. Vol. 3, p. 143.

STOUT, A. P., and KENNEY, F. R. (1949) Primary plasma cell tumours of upper air passages and oral cavity. *Cancer*, **2,** 261.

WEBB, H. E., HARRISON, E. G., MASSON, J. K., and REMINE, W. H. (1962) Solitary extramedullary myeloma (plasmacytoma) of the upper part of the respiratory tract and oropharynx. *Cancer*, **15,** 1142.

WILLIS, R. A. (1941) Solitary plasmacytoma of bone. *Journal of Pathology*, **53,** 77.

WILTSHAW, E. (1971) Extramedullary plasmacytomas. *British Medical Journal*, **2,** 327.

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