The Journal of Laryngology and Otology
July 1991, Vol. **105**, pp. 568–570

Clinical Records

Malakoplakia of the temporal bone in a nine-month-old infant

R. C. NAYAR,* I. GARG,** J. J. ALAPATT* (Bangalore, India)

Abstract
A case of malakoplakia, of the temporal bone in a nine-month-old male child is reported.

The lesion presented as an aural polyp, associated with a lower motor neuron facial palsy. On exploration, the granuloma was noted to involve the temporal bone, eroding the bony labyrinth. It was successfully treated with surgical debridement, and antibiotics. A review of the relevant literature is presented.

Introduction

Malakoplakia is the result of an unusual and distinctive type of inflammatory response, originally described in 1903, by von Hansemann in the urinary bladder, but subsequently reported in most other organs of the human body (Damjanov and Katz, 1981). In the otolaryngological literature malakoplakic lesions have been described in the palatine tonsils, (Kalfayan and Seager, 1982), middle ear (Azadeh and Ardehali, 1983), maxillary sinus (Nistal *et al.* 1985), nasopharynx (McCormick and Timme, 1982), base of tongue (Love *et al.*, 1985), larynx (Gabrielides *et al.*, 1981) and parotid gland (Dale and Robinson, 1988).

We present a case of malakoplakia, involving the temporal bone in a nine-month-old infant, highlighting interesting points in the presentation and pathology.

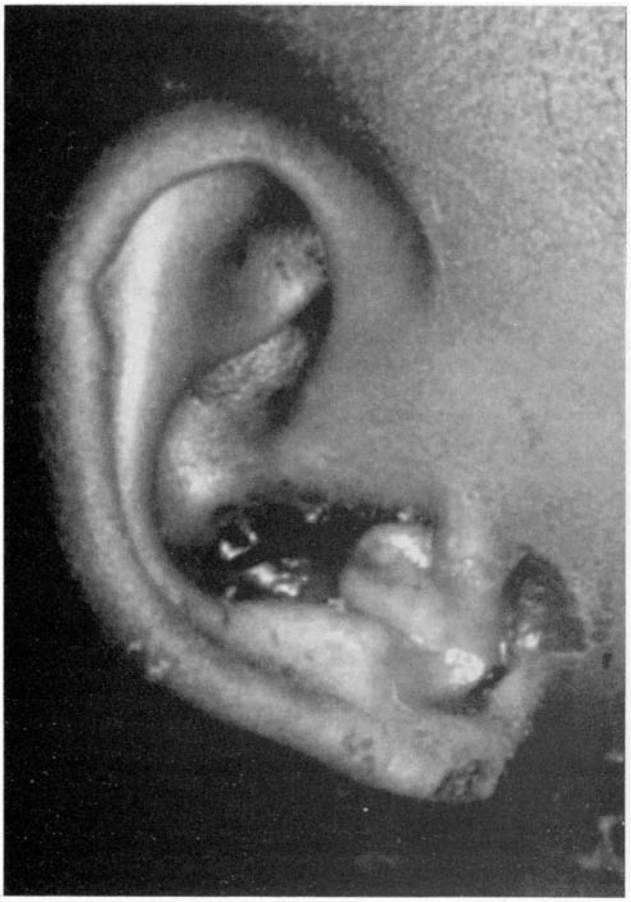

FIG. 1
Clinical photograph of malakoplakia presenting as an aural polyp.

Case report

A nine-month-old male child, first born of a non-consanguineous marriage, presented to the ENT outpatient depart-

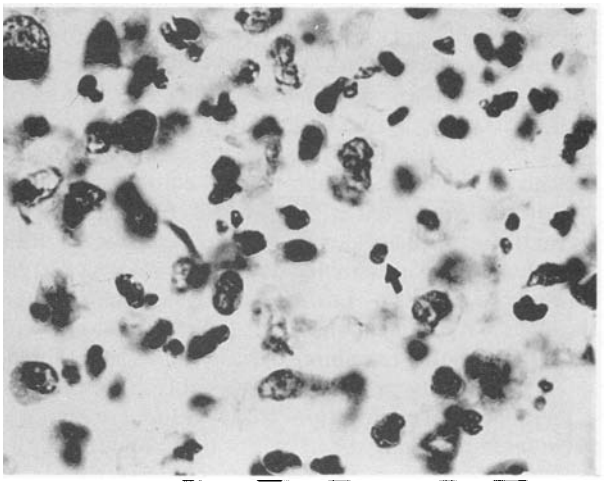

FIG. 2
Photomicrograph showing numerous macrophages and an extracellular laminated Michaelis-Gutmann body (H&E×100)

*Department of ENT, **Department of Pathology, St John's Medical College Hospital, Bangalore.
Accepted for publication: 18 March 1991.

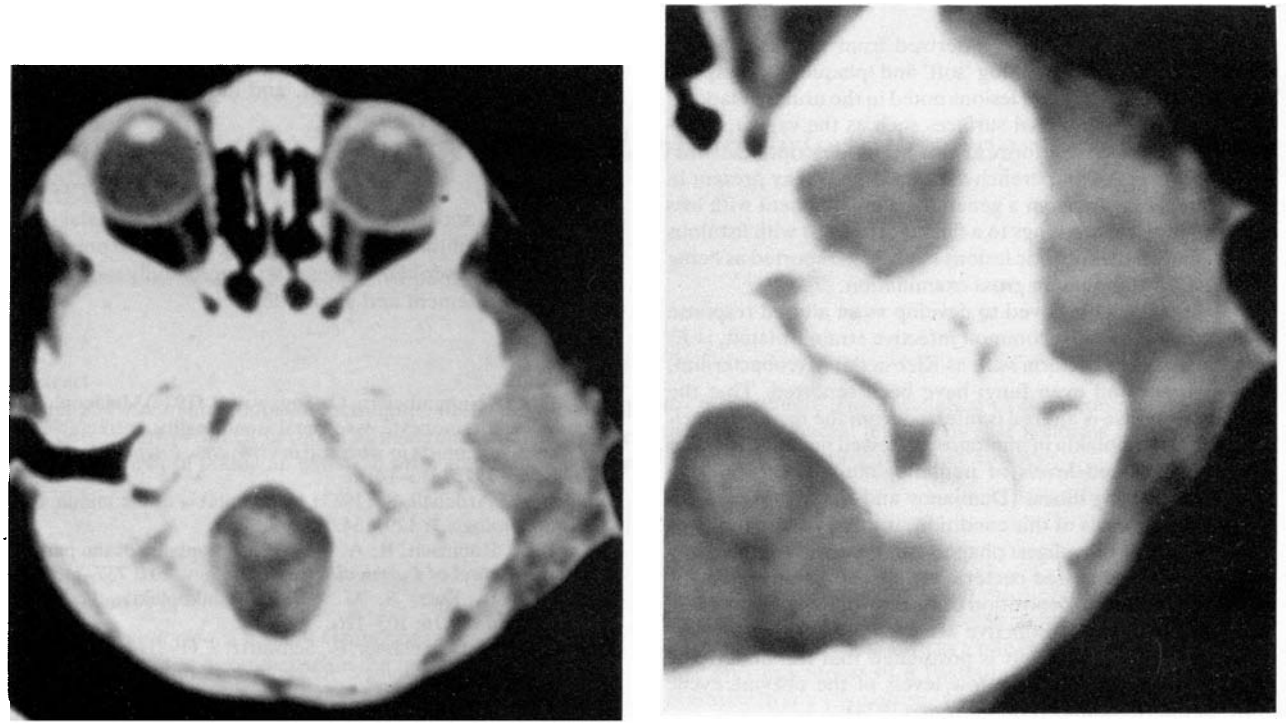

FIG. 3
CT scan showing lesion involving the right temporal bone and soft tissue extension.

ment of the St John's Medical College Hospital, Bangalore, in April 1990 with a mass in the right external auditory meatus and a post aural swelling, of four months duration. As the child was otherwise asymptomatic, these symptoms were ignored, until a progressive facial palsy on the same side was noted, a week prior to presentation.

On examination a papilliferous mass was seen in the right external auditory meatus. The pinna appeared to be raised by a swelling which extended from the parotid anteriorly obliterating the post-aural groove posteriorly. The margins of the swelling were ill-defined. Though the overlying skin was tense, there were no other signs of inflammation (Fig. 1). The facial palsy was of the lower motor neuron type.

Histopathology of the aural polyp revealed that the lesion was composed predominantly of histiocytes, though many plasma cells and lymphocytes were also present. Many histiocytes had eosinophilic granular cytoplasm. Calcosperites (Michaelis—Gutmann bodies) were present both within the histiocytes and extracellularly. These stained positive with PAS (for polysaccharides), Von Kossa (for calcium), and Perl's stain (for iron). These features were considered diagnostic of malakoplakia (Fig. 2).

A CT scan revealed a contrast enhancing lesion involving the right temporal bone, and soft tissues in the post-aural and parotid region, extending into the petrous apex, up to the clivus on the right side (Fig. 3).

Considering the large size of the lesion, a decision was taken to surgically debulk the tumour. However, given its benign nature, no attempt was made to radically excise the temporal bone. The surgery entailed initial identification of the facial nerve in the parotid; this was then followed through the lesion up to the second genu (Fig. 4). The friable tumour was removed piecemeal. The lesion appeared to involve the entire temporal bone, distorting the normal anatomy, and eroding the bony labyrinth. The Fallopian canal, proximal to the geniculate ganglion was spared. The medial limit of dissection stopped short of the internal carotid artery in the petrous apex.

The child withstood the surgery well and there was minimal blood loss. Post-operative medication (trimethoprim sulphamethoxazole) was given to the child who has since been discharged and is being kept on a close follow-up. The mastoid cavity shows evidence of epithelization. However, the facial palsy so far shows no signs of recovery.

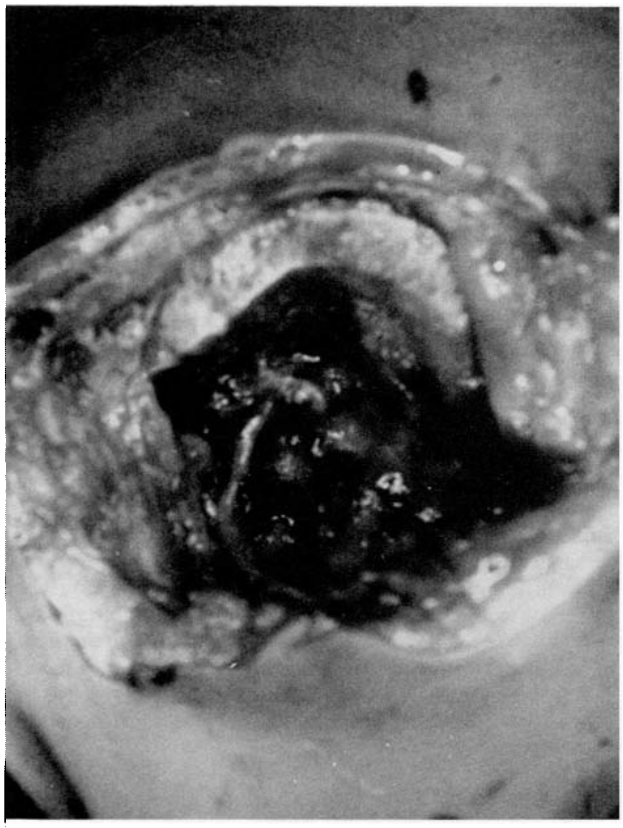

FIG. 4
Intra-operative photograph, showing the intact facial nerve, after surgical debridement.

https://doi.org/10.1017/S0022215100116627 Published online by Cambridge University Press

Discussion

The term 'malakoplakia' is derived from the Greek word '*malakos*' and '*plaks*' meaning 'soft' and 'plaque-like' respectively, describing the initial lesions noted in the urinary bladder. Lesions involving mucosal surfaces such as the vagina, intestines conjuctiva *etc* conform to this initial description. However, malakoplakia of parenchymatous organs may present in other forms ranging from a generalized enlargement with loss of morphological markings to a fluctuant abscess with fistulous tracts. Many malakoplakic lesions have been reported as being mistaken for tumours on gross examination.

Malakoplakia is believed to develop as an altered response to infection. The most common infective strain isolated, is *E. coli* though other bacteria such as Klebsiella, Mycobacterium, *Staph. aureus* and even fungi have been reported. That the immune response is altered is inferred from the increased incidence of malakoplakia in immunosuppressed patients, in those showing decreased levels of immunoglobulins, or suffering from a debilitating illness (Damjanov and Katz, 1981).

The pathogenesis of this condition, is due to the inability of macrophages to fully digest phagocytozed bacteria. Fragments of incompletely digested bacteria form the organic matrix for the intracytoplasmic deposition of calcium and other inorganic salts which form the distinctive Michaelis—Gutmann bodies. (Lou and Teplitz, 1974). It is postulated that the underlying biochemical defect is in the low levels of the enzyme cyclic GMP in the lysosomes (Abdou *et al.*, 1974).

This explains the therapeutic efficiency of cholinergic agonists (Bethanecol), which act by increasing the levels of cyclic GMP in the cell (Stauton *et al.*, 1983).

To the best of our knowledge, there is only one previously reported case of malakoplakia involving the middle ear (Azadeh and Ardehalli, 1983). The presenting symptoms in their patient were recurrent post-auricular abscesses and a lower motor neuron facial palsy. On exploration, extensive granulations were found in the mastoid cavity. The lesion was noted to have resolved satisfactorily with antibiotics. The authors stressed the importance of routine histopathological examinations of middle ear granulations for early diagnosis of malakoplakia.

Malakoplakia has been reported to occur in children, the youngest case being in the adrenal gland of a six-week-old male infant (Sinclair-Smith *et al.*, 1975). Gupta *et al.* (1972) reported a patient with multiple discrete lesions in the kidney, lung, lumbar and sacral vertebrae. The nature of the bone lesion as described in the report, is similar to that of the present case.

The present case is noteworthy in that the lesion occurred in a very young patient with no demonstrable evidence of malnourishment or tuberculosis. There was extensive destruction of the temporal bone including the bony labyrinth. Though a facial palsy was present with the lesion encompassing the facial nerve in the temporal bone, the nerve was demonstrably intact. Surgical exploration was considered necessary for debridement, given the large size of the lesion and to permit detailed histopathological examination of the excised specimen, as coexistence of malakoplakia with carcinoma has been described (McCormick and Timme, 1982).

Cholinergic agonists were not used, as the lesion regressed following surgical debridement, and the use of conventional antibiotics.

Conclusion

The clinical and pathological features of a case of malakoplakia in a nine-month-old child, presenting as an aural polyp with facial palsy, is described. The case was successfully treated with surgical debridement and antibiotics.

References

Abdou, N. I., Napombejara, C., Sagawa, A. (1974) Malakoplakia, evidence for moncyte lysosomal abnormality correctable by cholinergic agonists *in vitro* and *in vivo*. *New England Journal of Medicine,* **297:** 1413–1415.

Azadeh, B., Ardehali, S. (1983) Malakoplakia of the middle ear. *Histopathology,* **7:** 129–134.

Dale, J. C., Robinson, R. A. (1988) Malakoplakia of the parotid gland . *Journal of Laryngology and Otology,* **102:** 737–740.

Damjanov, I., Katz, S. M. (1981) Malakoplakia. *Pathology Annual, Part 2,* **16:** 103–126.

Gabrielides, G. G., Schimer, F., Schwartz, S. (1981) Malakoplakia of the larynx. *Pathology, Research and Practice,* **172:** (1–2) 53–57.

Gupta, R. K., Schuster, R. A., Christian, W. B. (1972) Autopsy findings in a unique case of malakoplakia. *Archives of Pathology,* **93:** 42–47.

Kalfayan, B., Seager, G. M. (1982) Malakoplakia of the palatine tonsil. *American Journal of Clinical Pathology,* **78:** 390–394.

Lou, T., Teplitz, C. (1974) Malakoplakia, pathogenesis and ultrastructural morphogenesis. A problem of altered macrophage response. *Human Pathology,* **5:** 191–195.

Love, B. B., Stanford, H. S., Jones, B. (1985) Malakoplakia of the tongue base. A case report. *Journal of Otolaryngology,* **14(3):** 179–182.

McCormick, K., Timme, A. H. (1982) Malakoplakia of the nasopharynx associated with carcinoma. *Journal of Laryngology and Otology,* **96:** 737–742.

Nistal, M., Regadera, J., Razquin, S., Paniagua, R., Gavilan, J. (1985) Malakoplakia of the maxillary sinus. *Annals of Otology, Rhinology and Laryngology,* **94:** 117–120.

Sinclair Smith, C., Kahn, L. B. N., Cywes, S. (1975) Malakoplakia in childhood, case report with ultrastructural observations and review of literature. *Archives of Pathology,* **99:** 198–203.

Stauton, M. T., Pearce, S. B., Maingot, P. P. (1983) Malakoplakia of the bladder treatment with trimethoprim, sulfamethoxazole, ascorbic acid and bethanechol. *Journal of Urology,* **13: 70(6):** 174–175.

Address for correspondence:
Dr Ravi C. Nayar,
Assistant Professor,
Department of ENT,
St John's Medical College Hospital,
Bangalore,
India 560034.

Key words: Malakoplakia; Temporal bone pathology