The Journal of Laryngology and Otology
June 1999, Vol. **113**, pp. 578–580

Pathology in Focus

Rapidly invading sebaceous carcinoma of the external auditory canal

J. RAY, M.S., F.R.C.S., J. B. SCHOFIELD, M.R.C.PATH.*, J. C. SHOTTON, F.R.C.S., A. AL-AYOUBI, M.D.

Abstract
A very rare case of a sebaceous carcinoma of the external auditory canal with basal cell differentiation is presented. Fewer than 400 cases affecting any part of the body have so far been reported and of that only seven cases have been known to involve the external auditory canal. The clinical features, pathology and treatment are described and the relevant literature has been reviewed.

Key words: Ear, external; Sebaceous gland neoplasms; Neoplasms, basal cell

Introduction

Cancer of the ear is quite uncommon and sebaceous cell tumours in that region are extremely rare. Fewer than 400 cases affecting any part of the body have so far been described (Bailet *et al.*, 1992). The commonest site is the upper eyelid. Extraocular sites include the parotid and submandibular glands, external genitalia, the trunk, extremities and the external auditory canal. Only seven cases affecting the external auditory canal have been reported.

A case is reported here where a 50-year-old female with sebaceous carcinoma involving the external auditory canal and middle ear was treated with extended radical mastoidectomy. Local recurrence was treated by radiotherapy but succumbed to massive local invasion within 15 months of presentation. A case is also made for the use of combination surgery and radiotherapy.

Case report

Presentation

A 50-year-old Asian lady presented at out-patients with a fleshy mass filling the right external auditory canal. There had been a long history of drum perforation since childhood with subsequent hearing loss and chronic ear discharge. During the preceding few months the right ear discharge had been incessant and blood-stained. There was no earache, headache, dizziness nor tinnitus. Cranial nerve function was intact and neck nodes were not palpable.

Investigations

The mass was biopsied in the clinic and histology showed a tumour composed of islands of sebaceous cells admixed with undifferentiated basaloid cells (Figure 1). In areas the tumour was deeply infiltrative and composed of islands almost entirely of undifferentiated basaloid type cells (Figure 2) with cytological atypia and frequent mitotic features (Figure 3). The differential diagnosis on the biopsy included sebaceous epithelioma and sebaceous carcinoma. High resolution computed tomography (CT) scan of the temporal bone revealed a soft tissue mass involving the right external auditory canal and extending into the middle ear and post-auricular sulcus. The bony landmarks were intact (Figure 4).

Within two weeks of presentation and following the biopsy the patient developed an irregular swelling in the right post-auricular region with worsening pain and discomfort.

Surgery

An extended radical mastoidectomy with removal of the cartilaginous external canal and pinna along with removal of the mastoid process and attached soft tissue was undertaken. The tegmen was intact although tumour filled the hypotympanum. A temporalis rotation flap was used to cover the defect. Facial nerve function was lost inadvertently in the course of the surgery. Nerve grafting was not undertaken and a lateral tarsorraphy was performed to protect the eye on the affected side. The tumour removed was irregular and about 3 cm in its greatest dimension. Margins of resection were clear of tumour. The patient made good post-operative recovery with good healing and disappearance of symptoms.

Recurrence

However, after three months the patient developed another soft tissue swelling near the anterior margin of the resection. No neck nodes were palpable. A fine needle aspiration cytology and a trucut biopsy suggested recurrence of the primary pathology. She was treated with a full dose of radiotherapy with reasonable control for four months without further spread or complications.

From the Departments of Otolaryngology and Pathology*, Kent County Ophthalmic and Aural Hospital, Maidstone, Kent, UK.
Accepted for publication: 12 March 1999.

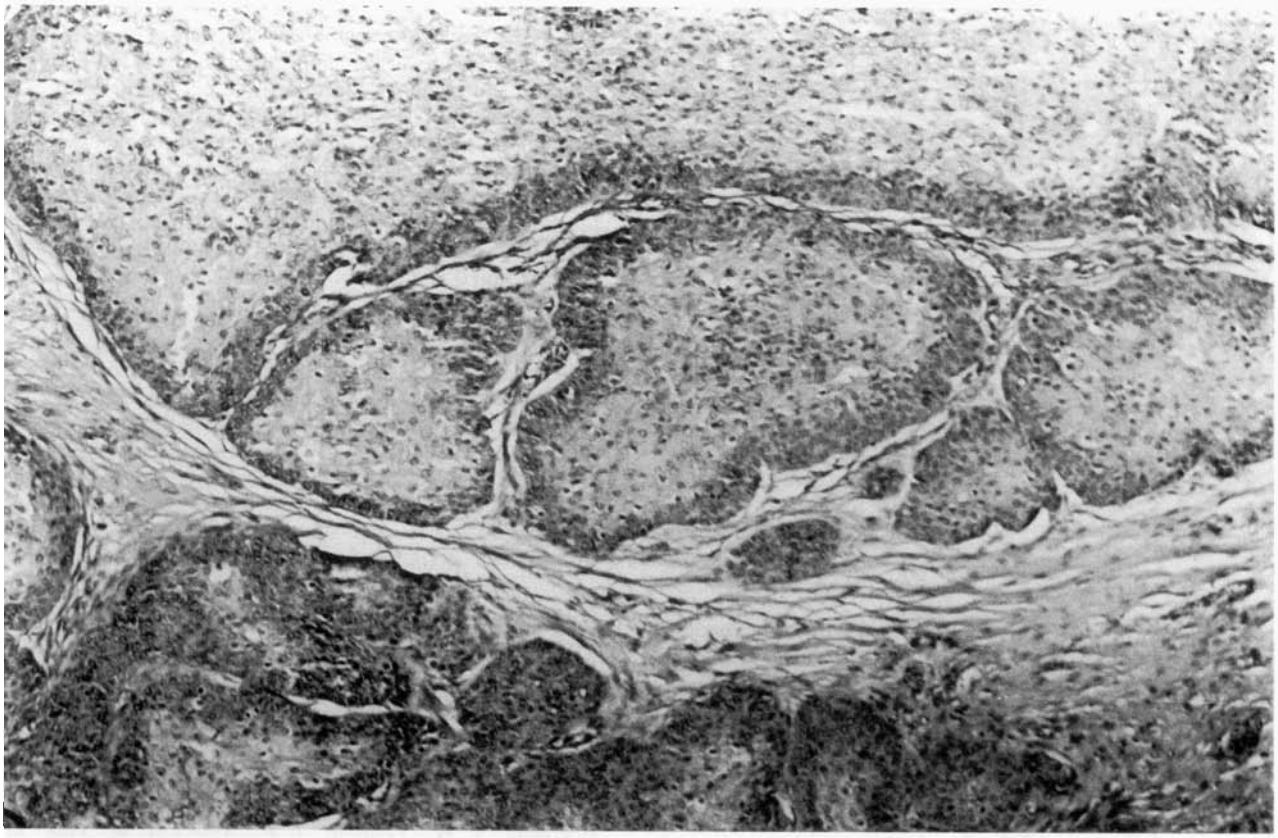

FIG. 1
Low power view showing a sebaceous tumour with a lobular architecture (H & E; × 140).

Further recurrence and spread

At this stage the patient became a non-attender for about seven months when she returned with widespread recurrence leading to skin breakdown and soft tissue infiltration. There was excruciating deep boring pain in the region. No neck nodes were palpable but the lower four cranial nerves were affected. Trismus was present. Spread to the skull base and infratemporal fossa with dural involvement rendered the disease inoperable. Palliative medical and nursing care was provided.

Outcome

She succumbed to the pathology from massive local infiltration and systemic infection 15 months after initial presentation.

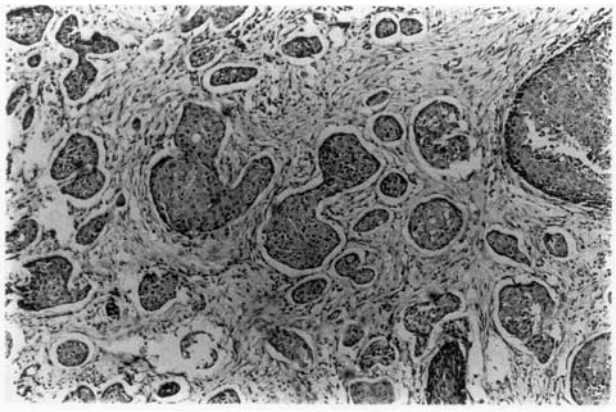

FIG. 2
High power view of section showing deeper layers of the tumour where there are smaller islands of sebaceous cells and a more infiltrative pattern associated with stromal desmoplasia (H & E; × 450)

Discussion

Incidence

Sebaceous carcinoma of the external auditory canal and middle ear are extremely rare and only seven cases have so far been reported (Doble *et al.*, 1981; Saito *et al.*, 1991; Bailet *et al.*, 1992).

Pathology

These tumours may basically be of three types: 1) sebaceous adenoma 2) basal cell carcinoma with sebaceous differentiation and 3) true sebaceous carcinoma (Rulon *et al.*, 1974). However there is a great degree of overlap between them and accurate nomenclature may be difficult (Verlooy *et al.*, 1993). Histological examination of

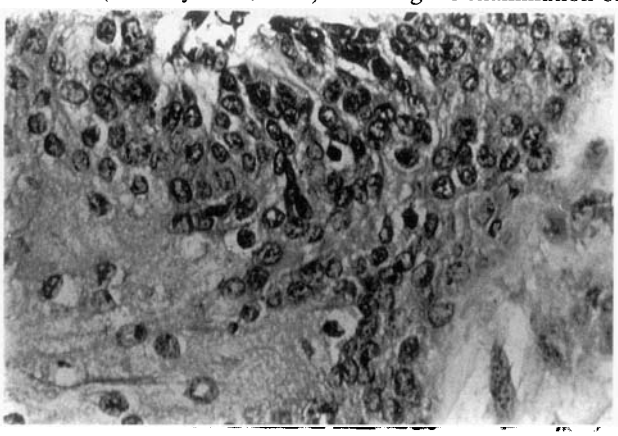

FIG. 3
High power view of section showing deep islands with marked cytological atypia and frequent mitotic figures (H & E; × 450)

https://doi.org/10.1017/S0022215100144536 Published online by Cambridge University Press

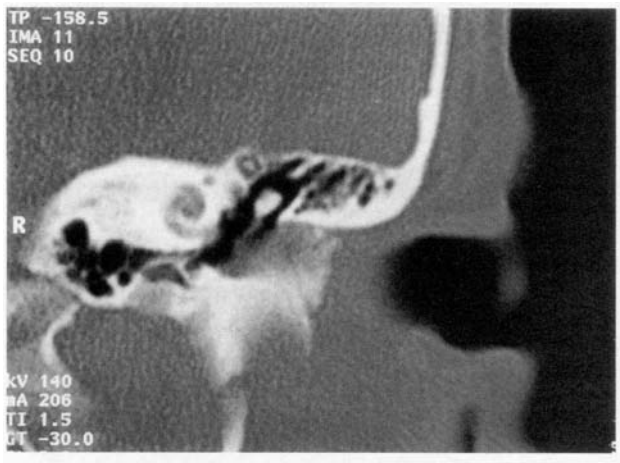

FIG. 4

Pre-operative high-resolution CT scan of the right temporal bone showing soft tissue tumour involving the external auditory canal and hypotympanum.

the specimen is the basis of diagnosis while fine needle aspiration cytology is useful in diagnosing recurrence and nodal spread (Bailet *et al.*, 1992).

It has been thought that extra-ocular sebaceous carcinomas are more likely to behave in a biologically aggressive manner and prognosis is poor (Wick *et al.*, 1985). Local infiltration and erosion are the commonest modes of spread but nodal and distant metastasis have been noted (Jensen, 1990). Involvement of lower cranial nerves, major blood vessels and dura points towards incurability. In our case this stage came within 15 months of initial presentation and the only option remaining was palliation.

Imaging

Pre-operative imaging is important both for knowing the extent of the tumour and also in planning treatment. High resolution computed tomography (HRCT) provided good contrast between osseous structures, air and soft tissues along with additional information about osseous defects and erosions (Figure 4). In most cases HRCT enables differentiation between inflammatory changes, cholesteatoma and tumour and is therefore the first imaging modality of choice for examining external and middle ear structures (Czerny *et al.*, 1997). However magnetic resonance (MR) imaging with or without infusion provides excellent delineation of soft tissue tumour margins, muscle infiltration and vascular encasement (Horowitz *et al.*, 1994).

Management

Radical surgery and radiotherapy preferably in combination are the accepted forms of treatment. However, the number of reported cases is too small to draw any statistically significant conclusion. In Stell's series (Stell, 1984) isolated local excision was sufficient. Others feel that a combination of surgery and radiotherapy is likely to give a better outcome (Lederman, 1965; Lewis, 1983). The rationale of this lies in debulking of the original tumour load, along with a large bulk of sclerotic bone, directing radiotherapy at residual disease. The drawback of single modality treatment is also exemplified in our case where the initial treatment modality was surgery. This led to an early and aggressive recurrence which subsequent radiotherapy failed to cure completely.

Conclusion

Sebaceous carcinoma of the ear is a very rare and highly aggressive tumour. The diagnosis is histological. The tumour is locally invasive and can also metastasize. Prompt radical surgery and radiotherapy in combination and in that order is the recommended treatment.

References

Bailet, J. W., Zimmerman, M. C., Arnstein, D. P., Wollman, J. S., Mickel, R. A. (1992) Sebaceous carcinoma of the head and neck-case report and literature review. *Archives of Otolaryngology – Head and Neck Surgery* **118(11):** 1245–1249.

Czerny, C., Turetschek, K., Duman, M., Imhof, H. (1997) Imaging of the middle ear. CT and MRI. *Radiologe* **37(12):** 945–953.

Doble, H. P., Snyder, G. C., Carpenter, R. K. (1981) Sebaceous cell carcinoma of the external auditory canal. *Journal of Laryngology and Otology* **68:** 114–119.

Horowitz, S. W., Leonetti, J. P., Azar-Kia, B., Fine, M., Izquierdo, R. (1994) CT and MR of temporal bone malignancies primary and secondary to parotid carcinoma. *American Journal of Neuroradiology* **15(4):** 755–762.

Jensen, M. L. (1990) Extra ocular sebaceous carcinoma of the skin with visceral metastases. *Journal of Cutaneous Pathology* **17(2):** 117–121.

Lederman, M. (1965) Malignant tumours of the ear. *Journal of Laryngology and Otology* **79:** 85–119.

Lewis, J. S. (1983) Surgical management of tumours of the middle ear and mastoid. *Journal of Laryngology and Otology* **97:** 229–311.

Rulon, D. B., Hewig, E. B. (1974) Cutaneous sebaceous neoplasms. *Cancer* **33:** 82–102.

Saito, C., Nishioka, K., Watanabe, S., Masuda, Y., Taguchi, K. (1991) Sebaceous epithelioma in the external auditory meatus: a case report. *Hiroshima Journal of Medical Sciences* **40(2):** 79–81.

Stell, P. M. (1984) Carcinoma of the external auditory meatus and middle ear. *Clinical Otolaryngology* **9:** 291–299.

Verlooy, H., Martelmans, L., Schiepers, C., DeRoo, M. (1993) A rare case of sebaceous carcinoma with malignant characteristics. *Clinical Nuclear Medicine* **18(5):** 425–427.

Wick, M. R., Goellner, J. R., Wolfe, J. T., Su, W. P. (1985) Adnexal carcinoma of the ear II: extraocular sebaceous carcinoma. *Cancer* **56:** 1163–1172.

Address for correspondence:
Mr J. Ray, M.S., F.R.C.S.,
Flat 2, 89 Longfleet Road,
Poole BH15 2HP.