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Pathology in Focus

Frozen section diagnosis in a jugulo-tympanic paraganglioma

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Abstract
The light microscopy and immunohistochemical findings of a jugulo-tympanic paraganglioma occurring in a 29-year-old man are reported. Diagnostic difficulties from frozen sections are stressed and selected literature is briefly commented upon.

Key words: Paraganglioma; Frozen sections

Introduction

Jugulo-tympanic paraganglioma (JTP) is, although rare, the most frequent tumour in the middle ear (Brown, 1985; Hyams *et al.*, 1988). Its behaviour is unpredictable in some locally aggressive cases, and for others with metastatic ability have also been reported (Brown, 1985; Hyams *et al.*, 1988; Bitterman *et al.*, 1992). Although both histological and immunohistochemical (Warren *et al.*, 1985; Achilles *et al.*, 1991) patterns are usually well recognized, intraoperative diagnosis could eventually be troublesome. In fact, little has been written about how to recognize and differentiate this entity from frozen sections.

We report a new case of JTP enhancing its morphological peculiarities intraoperatively. Differential diagnosis with other highly vascularized lesions in the middle ear is briefly mentioned.

Case report

A 29-year-old man presented with hearing loss and tinnitus. His symptoms had begun five months earlier. Clinical exploration revealed a polyp protruding into the left external auditory canal. Radiological examination showed a tumour mass occupying the middle ear. Surgical resection with intraoperative histology was performed. Clinical diagnosis at surgical exploration included cholesterol granuloma, paraganglioma, and meningioma.

Pathological findings

A haemorrhagic specimen, 1 cm in diameter, was received for intraoperative histopathology. Frozen sections showed highly vascularized tissue. Fibrous and oedematous areas were seen. Cuboidal cells were arranged in solid cords and small nests. Careful search displayed a few organoid nests containing large cells with pleomorphic and hyperchromatic nuclei (Figure 1). Therefore, a diagnosis of JTP was cautiously offered to the surgeon. Additional surgical specimens, 1.5 × 0.5 × 0.5 cm altogether, were then submitted. Formalin-fixed and paraffin-embedded tissue cut sections showed that the neoplasm grew beneath the surface squamous epithelium. Small cellular nests and ribbons were lying in a fibrous stroma with a mild lymphocytic infiltrate present. The vascular network was prominent. A close examination displayed occasional large nests with orga-

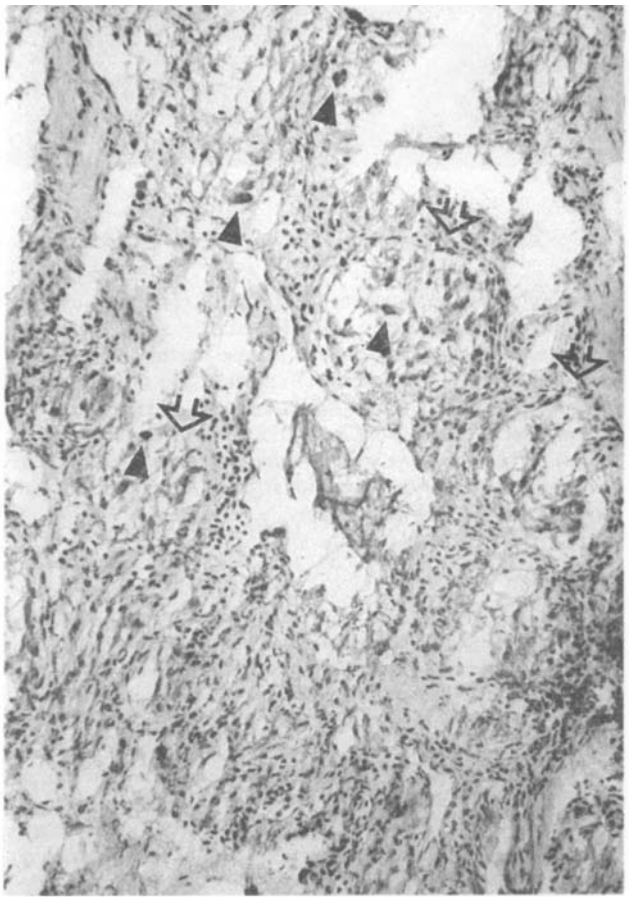

FIG. 1

Frozen section showing a cellular neoplasm displaying subtle nests with organoid (Zellballen) appearance (arrows) and occasional large cells with dark stained nuclei (arrowheads) (H&E × 250).

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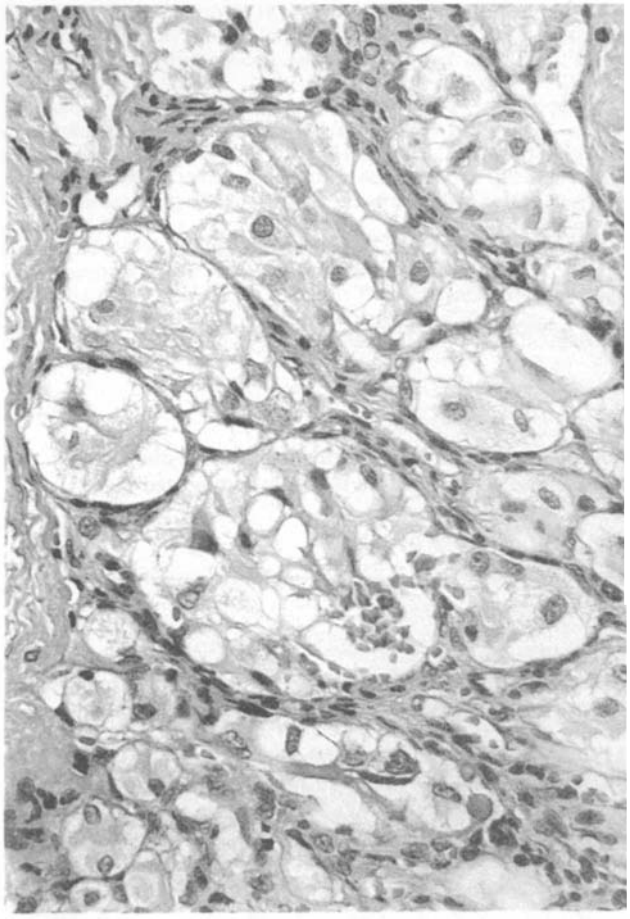

FIG. 2
Chief cells are arranged in organoid nests (Zellballen) (H&E × 400).

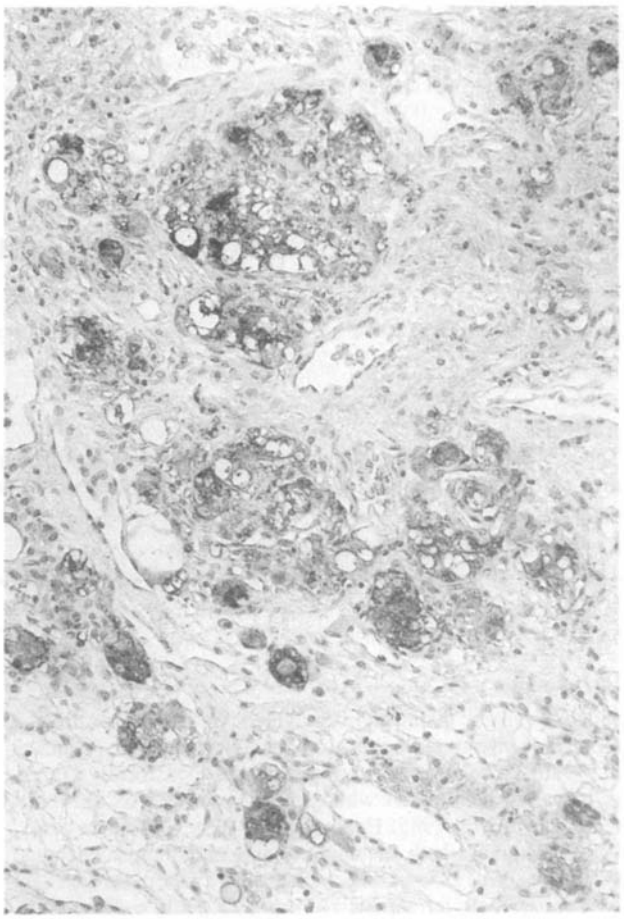

FIG. 3
Strong immunoreactivity for synaptophysin (× 250).

noid (Zellballen) arrangement of chief cells (Figure 2). Atypia was mild and mitoses were not observed.

Immunohistochemistry showed intense reaction with γ-enolase (Monosan), chromogranin (Biogenex) and synaptophysin (Biogenex) (Figure 3). AE1–AE3 keratin (Hybritech), Cam 5.2 keratin (Becton-Dickinson) and glial fibrillary acidic protein (Dako) all gave negative results. Very occasional sustentacular cells, not detected with conventional stains, were marked with antibody to S-100 protein (Dako).

Discussion

In normal conditions, frozen section diagnosis of a retroperitoneal paraganglioma should not be a difficult task (Silva and Balfour-Kraemer, 1987). However, rapid diagnosis of JTP can be difficult. This tumour contains less organoid nests than conventional paraganglioma and is often devoid of typical sustentacular and ganglion cells (Glenner and Grimley, 1974; Bitterman *et al.*, 1992). Prominent stromal fibrosis can be observed. Chief cells tend to be arranged in cords between the vascular network. Furthermore, small spindle cell variants have also been reported (Bitterman *et al.*, 1992). Current books on pathology have not stressed enough these peculiarities concerning intraoperative diagnosis (Glenner and Grimley, 1974; Silva and Balfour-Kraemer, 1987; Enzinger and Weiss, 1988; Hyams *et al.*, 1988).

To make the problem worse the middle ear can frequently harbour an aural polyp, non-specific granulation tissue, cholesterol granuloma, haemangioma, haemangiopericytoma, carcinoid, soft tissue alveolar sarcoma, schwannoma, meningioma, and epithelial neoplasm (Silva and Balfour-Kraemer, 1987; Enzinger and Weiss, 1988; Hymas *et al.*, 1988). It should be noticed that most of them are highly vascularized lesions. In addition they can eventually present fibrotic appearances and usually contain a dense inflammatory infiltrate.

All the aforesaid reasons contribute to make the diagnosis of JTP particularly difficult from intraoperative histology. The distinction between JTP and meningioma is perhaps the most problematic. The search for Zellballen structures and chief cells was definitive in securing an intraoperative diagnosis in the present case.

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