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Case report

Intravascular papillary endothelial hyperplasia of the skull base and intracranial compartment

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1. Introduction

Intravascular papillary endothelial hyperplasia (IPEH) was first described by Masson as a benign vascular tumor in a thrombosed hemorrhoidal vein in 1926 [4]. Histological and immunochemical analyses have since demonstrated IPEH to be a reactive proliferation of endothelial cells that can occur during thrombus organization [1,2,5]. In spite of its benign histology, IPEH may demonstrate aggressive behavior including progressive enlargement and recurrence after incomplete resection [2]. We present the clinical, imaging, and histological findings of three patients with IPEH involving the skull base and intracranial compartment.

2. Case reports

2.1. Case 1

This 56-year-old woman presented with chronic, recurrent sinus infections. Maxillofacial CT demonstrated an incidental 2.5 cm mass centered in the clivus with extension into the sphenoid sinus, right petrous apex, and right cavernous sinus. The clivus showed permeative erosion with elevation and thinning of the tuberculum sellae (Fig. 1A). MR imaging showed a lobulated, homogeneously enhancing, T2 hyperintense mass suspicious for possible chordoma or chondrosarcoma (Fig. 1B and C). The patient underwent trans-sphenoidal lesion biopsy, yielding tan-gray to white fragments of tissue. Mild blood loss was encountered during biopsy. Histology demonstrated a sclerotic vascular lesion with papillary fronds lined by a single layer of endothelial cells (Fig. 1D). Immunohistochemical analysis demonstrated cells positive for factor 8 and CD 31, confirming their endothelial nature. The cells were negative for AE1/3, S-100, D2-40, and EMA. These results were consistent with IPEH.

Given lesion location and potential morbidity of surgical resection, the patient was referred for stereotactic radiation therapy. However, radiation therapy was deferred due to the patient's lack of associated symptoms and she was followed with serial imaging. MR and CT imaging performed 10 months following biopsy demonstrated no significant change in lesion size or signal characteristics. The patient has remained asymptomatic now for nearly two years.

2.2. Case 2

This 39-year-old female originally presented with multiple syncopal episodes and was incidentally found to have a 1 cm lytic

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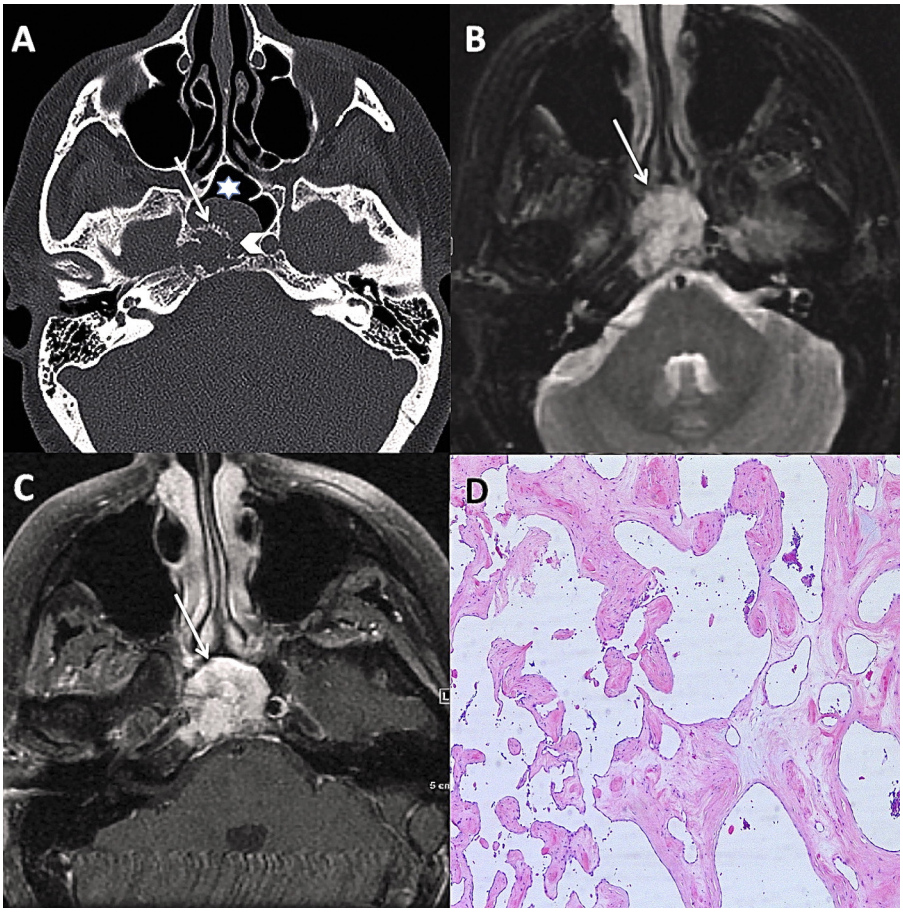

Fig. 1. 56-year old woman with skull base IPEH. (1A) Axial unenhanced head CT in bone window show an expansile, mass with areas of cortication suggesting a chronic process (arrow), as well as areas that appear more permeative and aggressive (arrowheads) involving the clivus and petrous apex, with exophytic extension into the sphenoid sinus (*) (1B,C) Axial T2- and post-contrast fat saturated T1-weighted imaging demonstrates a lobulated, homogenously enhancing, T2-hyperintense mass involving the central skull base (arrows). (1D) Representative hematoxylin and eosin micrograph demonstrates papillary structures lined by single layer of endothelial cells around a hypocellular fibrinous core within dilated vascular spaces (10x).

expansile lesion in the right petrous apex. She was conservatively managed for four years when follow up head CT demonstrated an increase in size of the lesion, with associated erosion of the right petrous apex and carotid canal and a few foci of peripheral calcification (Fig. 2A). On MRI, the lesion was predominantly hyperintense on T2, hypointense on T1, with avid contrast enhancement (Fig. 2B and C). The lesion extended into right Meckel's cave, abutted the posterior right petrous internal carotid artery, and bulged

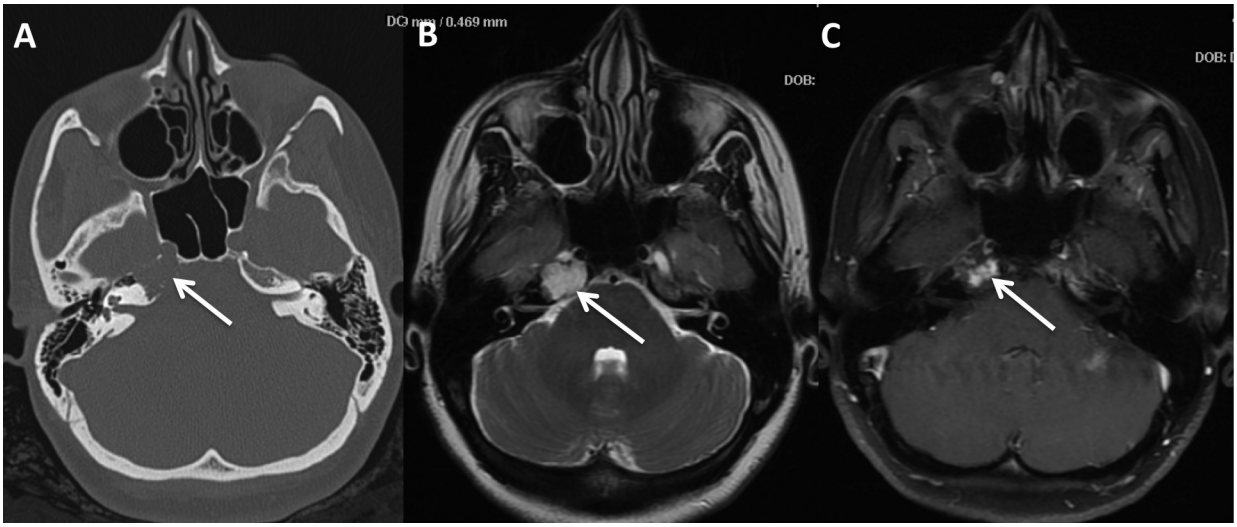

Fig. 2. 39-year-old female with skull base IPEH. (2A) Axial unenhanced head CT in bone window shows an expansile mass in the right petrous apex (arrow) with osseous erosion. (2B,C) Axial T2- and post-contrast fat saturated T1-weighted imaging demonstrates the mass to be heterogeneous, well demarcated, and avidly enhancing (arrows).