



A case of sinonasal paraganglioma with a different morphology: nine-year follow-up

Farklı morfolojiye sahip sinonasal paraganliyom olgusu: Dokuz yıllık takip

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ABSTRACT

Paragangliomas are very occasionally located in the nasal cavity. In this article, we present a 32-year-old female case with a sinonasal paraganglioma with atypical histopathological morphology. In our case, sinonasal paraganglioma consisted of lipoblast-like vacuolated cells instead of the typical Zellballen histological pattern. The prognosis of a nasal paraganglioma is not very clear. The present article is important, as it describes the first case report of a sinonasal paraganglioma with atypical lipoblast-like vacuolated cells with a nine-year follow-up.

Keywords: Follow-up; lipoblast-like vacuolated cell; nasal paraganglioma.

Öz

Paragangliyomlar çok nadiren burun boşluğunda yerleşimlidir. Bu yazıda, atipik histopatolojik morfolojiye sahip 32 yaşında sinonazal paragangliyomu olan bir kadın olgu sunuldu. Olgumuzda paragangliyom, tipik özelliği Zellballen histolojik paterni yerine lipoblast benzeri vakuollü hücrelerden oluşuyordu. Nazal paragangliyomların prognozu kesin olarak bilinmemektedir. Bu yazı, dokuz yıllık takip ile atipik lipoblast benzeri vakuollü hücrelerden oluşan ilk sinonazal paraganliyom olgusunu sunduğu için önemlidir.

Anahtar Sözcükler: Takip; lipoblast benzeri vakuollü hücre; nazal paraganliyom.

Paragangliomas are nonchromaffin neuroendocrine neoplasms that most commonly arise in adrenal glands, but they have also been recorded (5-10%) in extra-adrenal sites.^[1] Primary paragangliomas of the paranasal sinuses are very rare, with only a few cases described in the literature.^[2] Nasal paragangliomas are most commonly seen in middle-aged females in middle turbinate or ethmoid cells.^[2] Paragangliomas are locally aggressive, rarely recur, and can

metastasize.^[3] Therefore, complete resection and close follow-up are recommended.^[3,4]

In a typical histopathological examination of a paraganglioma, neoplastic or chief cells are arranged in well-defined nests, which have the classic alveolar or so-called zellballen pattern.^[5] An immunohistochemical study highlights the presence of S-100 protein-positive sustentacular cells at the periphery of the clusters of chief cells.^[6]



In this study, the chief cells were diffuse and showed intense positivity for neuron-specific enolase and synaptophysin.^[6]

We present a sinonasal paraganglioma case with atypical lipoblast-like vacuolated cells that was followed-up for nine years.

CASE REPORT

A 32-year-old woman presented with right-sided nasal obstruction, rhinorrhea, and a foreign body sensation for two years. Endoscopic nasal examination revealed a yellowish polypoid mass completely filling the right nasal cavity. The mass resembled an antrochoanal polyp. Paranasal computed tomography (CT) revealed a soft tissue mass filling the right nasal cavity and ethmoid sinuses (Figure 1).

Magnetic resonance imaging (MRI) confirmed an enhancing uniformly contrast medium mass, limited to the midline at the right lateral cranial site of the ethmoid bone and caudally extending below the middle nasal turbinate. There was no evidence of intracranial extension or bone destruction due to the mass (Figure 2).

An intraoperative frozen section biopsy revealed a neuroendocrine tumor. The mass originating from the right ethmoid roof was

completely excised endoscopically, with clear surgical margins. Frozen section examination revealed tumoral infiltration, which consisted of two cell types: small cells arranged in nests and the other one with large vacuolated cytoplasm. Paraffin-embedded tissue sections revealed polypoid structures lined with intact epithelia and a vascularized tumoral infiltration in the subepithelial region. The tumoral cells arranged in nests were small and had scant cytoplasm. The tumoral cells did not have the classic alveolar (zellballen) pattern. Lipoblast-like cells with vacuolated cytoplasm were also detected between the small cells. Immunohistochemistry revealed diffuse positivity for pancytokeratin, chromogranin, and synaptophysin, and S-100 was focally positive (Figure 3). No immunoreactivity was shown for vimentin, epithelial membrane antigen (EMA), mic-2 (CD99), or desmin. The Ki-67 proliferation index was 3-10%. According to the morphological features and immunohistochemical results, the diagnosis was a paraganglioma displaying lipoblast-like vacuolated cells.

The patient was followed with monthly endoscopic examinations for the first postoperative year and every three months thereafter. Control CT imaging was performed

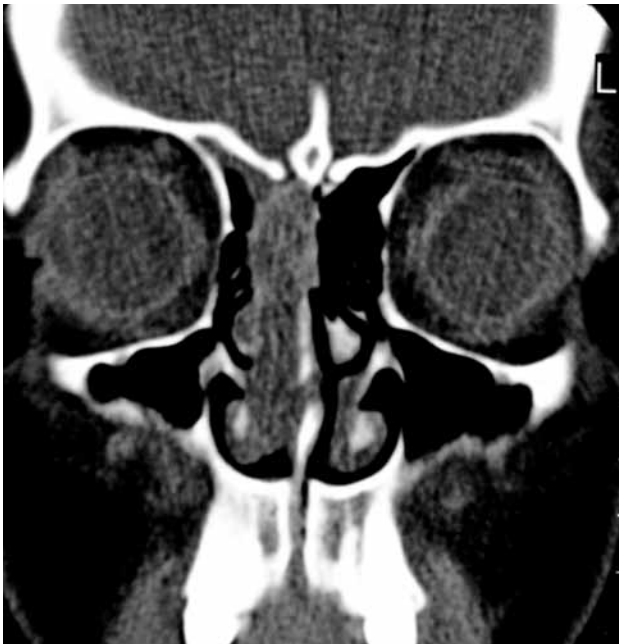


Figure 1. Paranasal sinus computed tomography, coronal section, showing soft tissue mass filling the right nasal cavity without bone erosion.

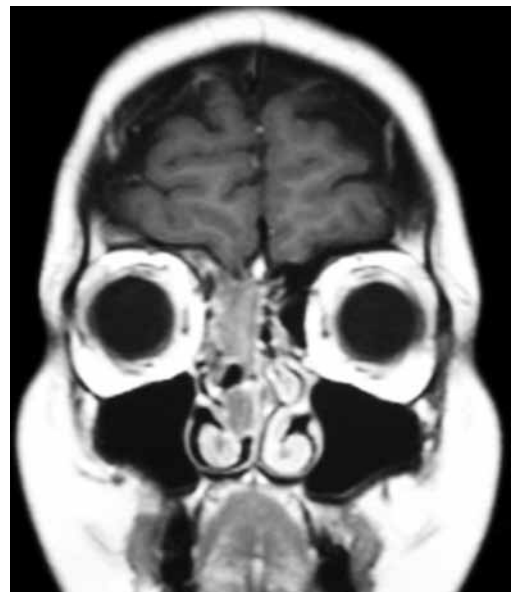


Figure 2. Magnetic resonance image confirming an enhancing uniformly contrast medium mass, limited to the midline at the right lateral cranial site of the ethmoid bone.

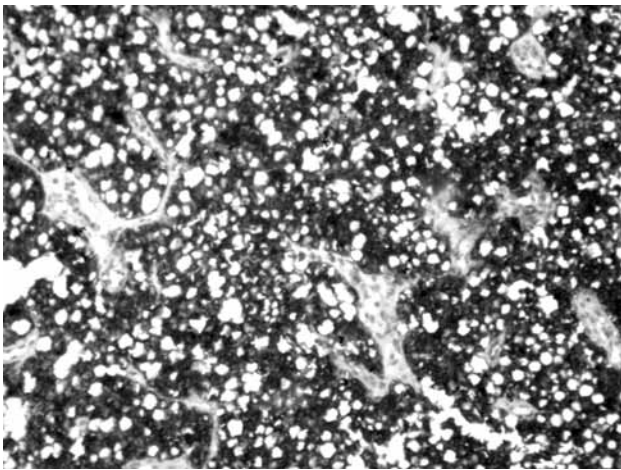


Figure 3. Immunohistochemical staining with pan cytokeratin, chromogranin, and synaptophysin, and S-100 (original magnification x 310).

every six months for the first two years after the surgery to check for recurrence. Magnetic resonance imaging was performed annually after the second year of the operation. The patient suffered from frontal headache three years after operation. An MRI scan revealed a 10x7 mm opacity at the frontal recess area. The mass and the mucosa of the frontal recess were resected endoscopically. The biopsy revealed a paraganglioma with the same morphology. One year after the second operation, the patient again suffered a unilateral headache, and CT and MRI showed another opacity in the frontal recess area. A Draf type 2 operation was performed, and the biopsy result was granulation tissue, with no evidence of any tumor. After the third operation, the patient was followed-up for five years with no evidence of tumoral recurrence or metastasis.

DISCUSSION

Sinonasal paragangliomas are slow-growing neoplasms, which may be present for months or years prior to detection.^[3] Paragangliomas occur over a wide age range (8-89 years), but the average age is 48.2 years.^[1] Females (as in our case) are more affected than males.^[1] Nasal paragangliomas develop mostly from the middle turbinate, lateral nasal wall, or the superior nasal vault.^[2,3] The most frequent symptoms are recurrent epistaxis, airway obstruction, and frontal headache.^[3,4] Complete surgical excision

is recommended because the tumor might be locally destructive.^[3] Our patient underwent surgery three times via an endoscopic approach.

The classic alveolar (zellballen) histological pattern seen in a typical histopathological examination of a paraganglioma was absent in our case. Instead, lipoblast-like vacuolated cells were seen on pathology. All previous sinonasal paragangliomas described in the literature had a typical zellballen pattern.^[1-6] To our knowledge, this is the first report of a sinonasal paraganglioma with atypical lipoblast-like vacuolated cells. The extraordinary morphology described herein is very rare. Tumors with such morphology may be difficult to diagnose, especially in the sinonasal region.

The precise prognosis of sinonasal paragangliomas is unknown, as only five sinonasal paraganglioma cases have been followed-up for more than eight years in the literature.^[1] Although our patient was followed-up for nine years, we still cannot generalize our results for others.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

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