Case 14865

Haemangiopericytoma of the oropharynx
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Section: Head & neck imaging
Area of Interest: Head and neck
Procedure: Diagnostic procedure
Imaging Technique: CT
Imaging Technique: MR
Special Focus: Neoplasia Case Type: Clinical Cases
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Patient: 60 years, male

Clinical History:
A 60-year-old man was admitted with a foreign body sensation in the throat associated with dysphagia and change in voice. He had no history of trauma or pain. Clinical examination revealed a large mass located on the left side of the oropharynx. Overlying mucosa appeared normal in colour.

Imaging Findings:
The computed tomography (CT) scan identified a large, well-marginated tumour of the oropharynx, arising from the left posterolateral wall - approximately at the level of the palatine tonsil, 5.5x3.8x5.3 cm in size, with intense peripheral enhancement. The lesion projects into the left parapharyngeal space, extending from the level of the medial pterygoid muscle to the hypopharynx and submandibular space. Magnetic resonance imaging (MRI) detected a large, soft-tissue mass of the oropharynx. On T1-weighted images the lesion reveals relatively homogeneous signal intensity similar to that of the muscles. On T2-weighted images and STIR images the mass displays heterogeneous hyperintensity with flow voids due to vessels in its internal structure and also internal areas of cystic changes, representing necrosis. On post-gadolinium images the tumour enhances intensively but with an inhomogeneous pattern. No lymphadenopathy was found on either of the two examinations. After surgical excision the histological evaluation characterised the lesion as HPC.

Discussion:
Haemangiopericytoma (HPC) is a very rare, soft tissue tumour of vascular origin formed by the proliferation of the pericytes of Zimmermann. These pericytes are modified smooth-muscle cells surrounding capillary vessels, located outside the reticulin sheath of the endothelium [1, 2]. Stout and Murray first described this tumour in 1942 [3]. As pericytes are present in the capillaries, HPC can occur anywhere in the body. Nevertheless, it is most commonly found in the trunk and the extremities. Less than 16% of the cases occur in the head and neck region, with cases reported arising in the orbit, nasal sinus tract, oral cavity, skull base, middle ear, buccal mucosa and larynx. Its occurrence in the pharynx is extremely rare.
HPS can occur in any age group. No sex predilection is noted. The biological-clinical behaviour and prognosis of HPC is unpredictable. According to the histological findings this tumour is classified as low-grade, intermediate-grade and high grade based on mitoses, cellularity, and cellular pleomorphism [2]. Enzinger and Smith suggested as malignant indices: tumour diameter >6.5 cm, mitotic figures >4 per high power fields, increased cellularity, cellular...
pleomorphism, necrosis and/or haemorrhage [4]. Nevertheless, the absence of necrosis, cellular pleomorphism, and mitoses <4 per 10 high power field does not necessarily indicate benignancy, since tumours with benign histologic appearance have been reported to metastasise [2, 5]. Moreover, a benign form of haemangiopericytoma does not exclude the possibility of recurrence years and even decades after surgical excision.

Since the vascular pattern expressed by such tumours is common to other tumours, the diagnosis of HPC is based on the following criteria: on immunohistochemical analysis, the HPC cells are negative for a-smooth muscle actin, desmin, S-100 protein, and cytokeratin, are intensely positive for vimentin, and focally positive for CD34 [2]. CT scan, Ultrasound, Doppler sonography and arteriography can provide further information regarding the location and the extent of tumor invasion into the surrounding tissue.

Surgical excision of the HPC lesion with preceding ligation of the vascular bundle that nourishes the tumour is the treatment of choice. Radiotherapy as adjuvant treatment is controversial since the tumour is considered to be radioresistant. However, it can be useful in cases of incomplete resections. Chemotherapy and immunotherapy can be used for control of malignant HPC and metastatic disease. Use of the CO2 laser technique can achieve both excision and cauterisation, and may thereby decrease the likelihood of local relapse and metastasis [5].

**Differential Diagnosis List:** Haemangiopericytoma of the oropharynx, Solitary fibrous tumour, Synovial sarcoma

**Final Diagnosis:** Haemangiopericytoma of the oropharynx

**References:**


Description: A large mass with well-defined margins arises from the posterolateral wall of the oropharynx. The mass shows heterogeneous, intense enhancement. Origin: Radiology Department of Papageorgiou General Hospital of Thessaloniki, Greece
Figure 2

Description: The tumour compresses the left parapharyngeal space and protrudes into the hypopharynx causing significant narrowing of the aerodigestive tract. Origin: Radiology Department of Papageorgiou General Hospital of Thessaloniki, Greece
Description: On T1W imaging the lesion reveals relatively homogeneous signal intensity similar to that of muscle. Origin: Radiology Department of Papageorgiou General Hospital of Thessaloniki, Greece
Figure 4

**Description:** The mass enhances avidly apart from necrotic areas. Note intact overlying mucosa. **Origin:** Radiology Department of Papageorgiou General Hospital of Thessaloniki, Greece
**Description:** The mass displays heterogeneous hyperintensity with flow voids due to vessels. High signal represents areas of cystic necrosis. **Origin:** Radiology Department of Papageorgiou General Hospital of Thessaloniki, Greece
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