Myopericytoma: a rare case of parapharyngeal localization

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ABSTRACT

This case study presents a 66-year-old man referred to the Otolaryngology and Head and Neck Surgery department due to a one-history of persistent pain in the left posterior cervical region. No abnormalities were detected in the oral and pharyngeal regions during clinical and endoscopic examinations. Subsequently, a magnetic resonance imaging revealed a lesion (14 x 12 x 14 mm) into the left parapharyngeal space, with high signal intensity on T2-weighted images, enhancement after contrast medium, restricted signal on diffusion weighted imaging and high vascularization on perfusion MRI. The histological examination of the lesion led to a diagnosis of myopericitoma. Post-surgery, no adjuvant therapy was administered. Myopericytomas are rare soft-tissue benign neoplasms, predominantly reported in extremities, with a limited number of cases in the head and neck region and almost never described in the literature with elective localization in the parapharyngeal space.

CASE REPORT

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A 66-year-old man was referred to the Otolaryngology and Head and Neck Surgery department. Pain in the cervical region was the patient's only symptom for about one year.

No alterations were demonstrated into the oral cavity, nasopharynx, oropharynx, hypopharynx and larynx during clinical and endoscopic examination using a flexible optical fiberscope and narrow band imaging (NBI) technology.

No palpable locoregional lymph nodes were present.

A contrast-enhanced 1.5T magnetic resonance imaging (MRI) was performed and it showed a non-infiltrating lesion of 14 (AP) x 12 (T) x 14 (L) mm, with an expansive growth and capsulated appearance, characterized by hyperintense signal on T2-weighted images, isointense to the muscle tissue on T1-weighted images, with low reduction in diffusivity on the diffusion weighted imaging (DWI) and mean values of 2.1x 10- 6 mm^2/s in apparent diffusion coefficient (ADC) maps; after contrast medium administration (Gadolinium 1 ml/ kg) an early, homogeneous and progressive intralesional post-contrast enhancement with poor and late washout was observed; perfusion MRI with Dynamic Contrast Enhanced (DCE) showed high blood and flow volume as a vascularized lesion (Figure 1).

The patient underwent surgical resection under general anesthesia after warning him about risks and having obtained his informed consent. The gross evaluation of the surgical specimen showed a well circumscribed nodule with rubbery texture and white-tan cut surface, measuring cm 1.3 in diameter. Microscopic examination revealed a well-defined and highly cellular lesion composed of oval to spindle-shaped cells with eosinophilic cytoplasm, with concentric growth around numerous thin and thick-walled blood vessels, variably sized. The stroma included focal areas of hyalinization and there was no evidence of necrosis, atypia or mitotic activity. Staining of the lesion showed strong positivity for smooth muscle actin (SMA) and h-caldesmon, while there was no expression of CD34. The morphologic features and immunohistochemical results were consistent with the diagnosis of myopericytoma (Figure 2).

The patient will be subjected to follow-up in the next few months with clinical examination and MRI in order to evaluate the presence of residual or radiological recurrence of the disease.

DISCUSSION

Etiology & demographics

Myopericytomas are uncommon, almost always benign and slow-growing soft-tissue neoplasms. The term is used

to describe tumors with a striking concentric perivascular proliferation of myoid differentiated pericytic cells [1], which generally arise within the dermis and subcutaneous tissue of the extremities. It is a rare finding in the head and neck, and even rarer is its deep location [2]: some recent literature reviews such as that of Roig et al. and Prado Calleros et al., identify a small number of lesions mostly localized in the parotid space and oral mucosa [3,4]. In particular, a review of the literature, performed using the MESH terms "(myopericytoma) AND ((head and neck) OR (oral) OR (parapharyngeal))" carried out using the main search engines for scientific articles such as PubMed and Google Scholar, identified a single study conducted on a myopericytoma localized in the parapharyngeal space [2].

Clinical & imaging findings

The diagnosis of myopericytoma is very difficult due to the small number of patients in whom this type of neoformation occurs. Patients with neoplasms of the parapharyngeal space such as myopericytoma usually present a single slow-growing and painless swelling or mass in the absence of cervical lymphadenopathy or other signs of neurological impairment, and without concomitant oncological history [5]. Fiber optic laryngoscopy does not reveal any further alterations of pathological significance.

The few data present in the literature about the radiological appearance of myopericytoma describe a lesion with different morphological and radiological characteristics, with or without sharp margins, described in most of cases like a non-infiltrating, heterogeneous mass, and with heterogeneous enhancement after intravenous injection of contrast medium [2–5]. These data are in partial disagreement with our experience, which instead demonstrated a homogeneous mass and post-contrast enhancement. In our case the diffusion MRI showed low restricted signal in relation to low cellular component, while the perfusion MRI showed a vascularized lesion with high blood and flow volume. In related studies on ultrasound data, the inhomogeneous appearance of the lesion is also confirmed, showing marked internal vascularity at Color-Doppler sonography, while no significant data are available regarding the evaluation with fluorodeoxyglucose-positron emission tomography (18F-FDG-PET) examination.

The treatment of this type of parapharyngeal space lesion is purely surgical. Local recurrence and malignancy have been observed only in a few cases in myopericytoma. The major but still rare complication in this type of surgery is blood loss due to the rich vascularization of these lesions and their growth adjacent to vascular structures [6].

Fine-needle biopsy in most of cases has not sufficient accuracy due to insufficient quantity of tissue sample [5].

Myopericytomas are well-defined and unencapsulated nodular lesions, characterized by variable cellularity and composed of oval to spindle-shaped myoid cells arranged around numerous vessels, which may be small and rounded or branching and ectatic [6].

By immunohistochemistry, myopericytoma's cells express SMA and h-caldesmon, whereas they are typically negative or only focally positive for desmin; immunostaining for CD34 is usually negative and it highlights the prominent vascular component.

Very rare examples of malignant myopericytoma have been reported [7], defined by marked nuclear atypia, increased proliferative activity and a deeply infiltrative growth.

DIFFERENTIAL DIAGNOSIS

Schwannoma and Paraganglioma

Schwannomas of the parapharyngeal space usually occur in patients aged between 30 and 70 years. Although schwannomas are typically benign, they can affect adjacent tissues resulting in a mass effect with dislocation of the adjacent vascular and nervous structures. On MRI, schwannomas are well-circumscribed homogeneous masses that exhibit high signal intensity on T2-weighted images and relatively homogeneous low signal intensity on T1-weighted images with moderate enhancement after contrast medium injection. Similar MRI features can be recognized in parapharyngeal space's paragangliomas. However, differently from paragangliomas, no vascular flow voids are observed in schwannomas [8,9].

Mucoepidermoid carcinoma

Mucoepidermoid carcinoma (MEC) is the most common malignant tumor of the salivary glands and accounts for 3-15% of all salivary gland cancers. The preferred site is the parotid gland, accounting for more than 80% of MECs. On MRI, MECs show uneven signal intensity on T2-weighted images, low to intermediate depending on histologic grade, and have an ill-defined margin [10].

Adenoid cystic carcinoma

Adenoid cystic carcinoma (ACC) occurs most frequently (60%) in the minor salivary glands distributed across the mucosa of the head and neck; the most common sites are the palate, tongue, sinuses, and nasal cavity.

These lesions usually show hypo-isointense signal in T1-weighted images, slightly hyperintense signal in T2-weighted images, with the exception of highest degrees markedly hypointense, and homogeneous post-contrast enhancement after contrast medium administration [11,12].

Pleomorphic adenoma

Pleomorphic adenomas (PAs) are the most common salivary gland tumor. In most cases they are located in the parotid gland, approximately 80% in the superficial lobe while only 20% in the deep lobe. PAs usually appear like well-defined encapsulated

lesions with smooth margins. On MRI, PAs are typically iso- or slightly hypointense compared to the surrounding muscle tissue on T1-weighted images, hyperintense on T2-weighted images and show heterogeneous enhancement after contrast medium injection. Cystic spaces or areas with different signal intensities can be seen within the tumor. These radiological features reflect the variety of histological components of the pleomorphic adenoma, representing a mixture of different tissues including epithelial, myoepithelial and stromal components. Sometimes, PAs may extend into the parapharyngeal space, infratemporal fossa or other adjacent regions [13,14].

Second branchial cleft cyst

Branchial cleft cysts (BCCs) are congenital malformations resulting from incomplete branchial remnants involution. Approximately 90%-95% represent a second branchial cleft abnormalities that usually become clinically evident between the ages of 20 and 40.

Type IV cysts are very rare and located in the pharyngeal mucosal space deep from the palatine tonsil and medial to the great vessels of the neck.

MRI usually shows a well-marginal mass, without postcontrast enhancement, which on T2-weighted images is slightly hyperintense compared to cerebrospinal fluid or hypointense if it contains pus [15].

TEACHING POINT

The masses of the parapharyngeal space are difficult to distinguish on the basis of clinical and radiological data, due to a large overlap of the findings: although the combination of morphological evaluation and the perfusion study can sometimes help to distinguish between benign and malignant pathology, it does not always manage to provide reliable answers. Because of this the surgical approach is recommended as the first choice and the anatomopathological analysis is the only way to confirm a diagnostic hypothesis and guide the clinical and therapeutic path. In consideration of what has been demonstrated in our experience, it is also necessary to consider myopericytoma among the differential diagnoses.

REFERENCES

[1] Granter SR, Badizadegan K, Fletcher CD. Myofibromatosis in adults, glomangiopericytoma, and myopericytoma: A spectrum of tumors showing perivascular myoid differentiation. *Am J Surg Pathol.* 1998; 22(5): 513-525. PMID: 9591720.

[2] Chu ZG, Yu JQ, Yang ZG, et al. Myopericytoma involving the parotid gland as depicted on multidetector CT. *Korean J Radiol*. 2009; 10(4): 398-401. PMID: 19568469.

[3] Roig NJ, Wu M, Hernandez O, Liu CZ, Brandler TC. Myopericytoma of the Parotid and Molecular Profiling: Report of a Rare Case and Review of the Literature. *Int J Surg Pathol.* 2022; 30(5): 574-580. PMID: 34970937.

[4] Prado–Calleros HM, Galarza–Lozano D, Arrieta–Gómez JR, et al. Myopericytoma arising adjacent to the common carotid artery: Case report and systematic review of deep located neck myopericytomas. *Head Neck.* 2016; 38(9): E2479-482. PMID: 27061437.

[5] Ju WT, Zhao TC, Liu Y, et al. Clinical and pathologic analysis of myopericytoma in the oral and maxillofacial region. *Oral Surg Oral Med Oral Pathol Oral Radiol.* 2019; 128(4): 393-399. PMID: 31350225.

[6] Mentzel T, Dei Tos AP, Sapi Z, Kutzner H. Myopericytoma of skin and soft tissues: Clinicopathologic and immunohistochemical study of 54 cases. *Am J Surg Pathol.* . 2006; 30(1): 104-113. PMID: 16330949.

[7] McMenamin ME, Fletcher CD. Malignant myopericytoma: Expanding the spectrum of tumours with myopericytic differentiation. *Histopathology.* 2002; 41(5): 450-460. PMID: 12405913.

[8] Miller FR, Wanamaker JR, Lavertu P, Wood BG. Magnetic resonance imaging and the management of parapharyngeal space tumors. *Head Neck.* 1996; 18(1): 67-77. PMID: 8774924.

[9] Wax MK, Shiley SG, Robinson JL, Weissman JL. Cervical sympathetic chain schwannoma. *Laryngoscope.* 2004; 32(2): 133–136. PMID: 22767977.

[10] Peraza A, Gómez R, Beltran J, Amarista FJ. Mucoepidermoid carcinoma. An update and review of the literature. *J Stomatol Oral Maxillofac Surg.* 2020; 121(6): 713-720. PMID: 32565266.

[11] Singh FM, Mak SY, Bonington SC. Patterns of spread of head and neck adenoid cystic carcinoma. Clin Radiol 2015;70. https://doi.org/10.1016/j.crad.2015.01.013.

[12] Hiyama T, Kuno H, Sekiya K, Oda S, Kobayashi T. Imaging of malignant minor salivary gland tumors of the head and neck. *Radiographics.* 2021; 41(1): 175-191. PMID: 33245669.

[13] Shin JH, Lee HK, Kim SY, Choi CG, Suh DC. Imaging of parapharyngeal space lesions: Focus on the prestyloid compartment. *AJR Am J Roentgenol*. 2001; 177(6): 1465-1470. PMID: 11717108.

[14] Almeslet AS. Pleomorphic adenoma: A systematic review. *Int J Clin Pediatr Dent.* 2020; 13(3): 284-287. PMID: 32904077.

[15] Lev S, Lev MH. Imaging of cystic lesions. *Radiol Clin North Am.* 2000; 38(5): 1013-1027. PMID: 11054966.

FIGURES

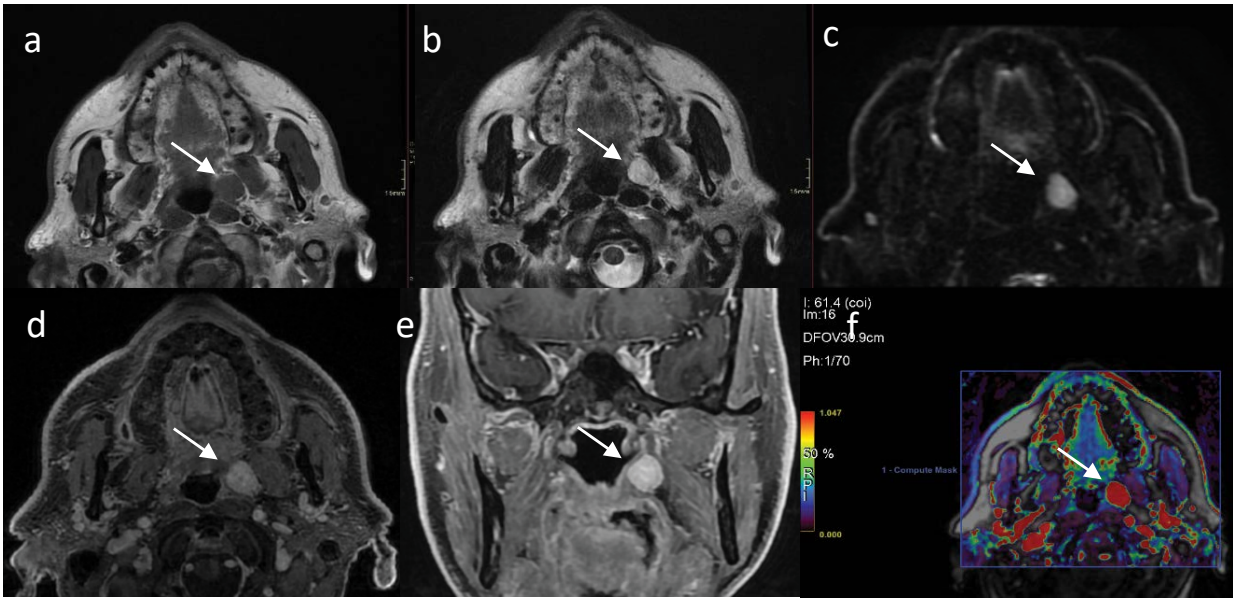

Figure 1: MRI of 66-year-old man. Findings: MRI shows a lesion into the left parapharyngeal space with an expansive growth, with isointense signal on T1 to muscle, hyperintense on T2, with restricted signal on DWI, homogeneous enhancement after contrast medium and high blood and flow volume on perfusion imaging. Technique: MRI at 1.5T, axial plane T1 sequence (a), axial plane T2 sequence (b), axial plane DWI with b 800 (c), axial and coronal planes T1 sequences with fat-suppressed after contrast medium (d-e) and perfusion sequence with Dynamic Contrast Enhanced (blood and flow volume – nIAUGC) (f).

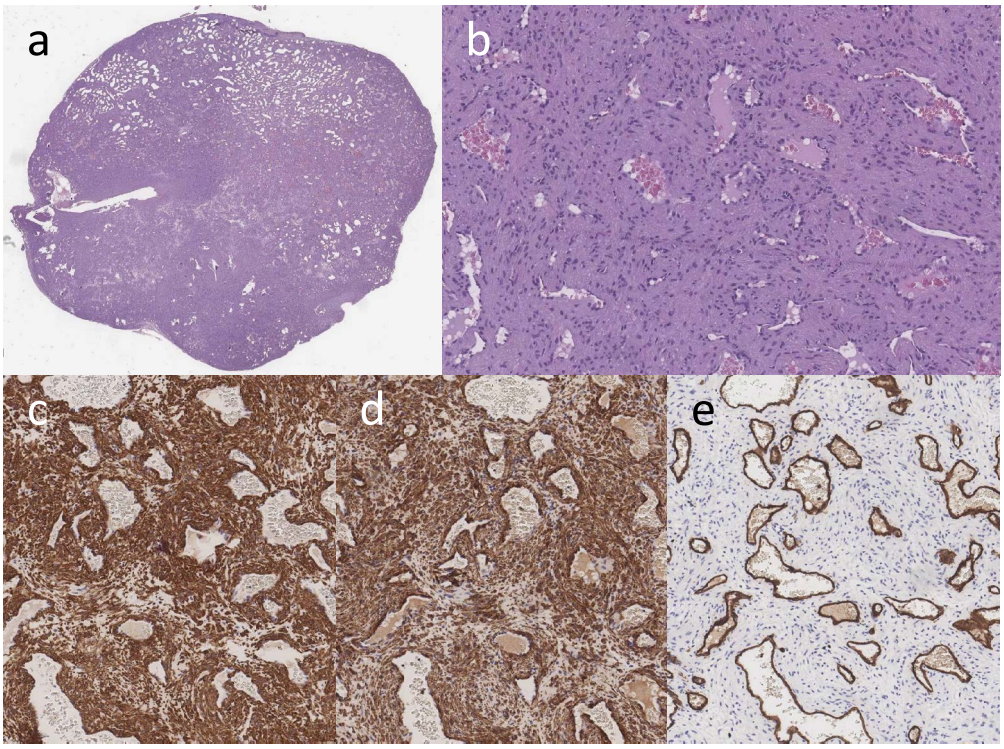

Figure 2: Histopathology examination: photomicrograph of the specimen showing a well-circumscribed neoplasm with numerous ectatic blood vessels (H&E, 10x) (a); photomicrograph of the specimen showing spindle-shaped cells with concentric growth around vascular channels (H&E, 100x) (b); SMA immunostaining, 100x (c); h-caldesmon immunostaining, 100x (d); CD34 immunostaining, 100x (e).

KEYWORDS

MRI, Myopericytomas, Parapharyngeal Space, Perfusion Imaging, Diffusion Imaging

ABBREVIATIONS

ACC = adenoid cystic carcinoma; ADC = apparent diffusion coefficient; BCCs = branchial cleft cysts; CT = computed tomography; DCE = dynamic contrast enhanced; DWI = diffusion weighted Imaging; MEC = mucoepidermoid carcinoma; MRI = magnetic resonance imaging; NBI = narrow band imaging; PA = pleomorphic adenoma; SMA = smooth muscle actin; 18F-FDG-PET = fluorodeoxyglucose-positron emission tomography

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