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Case Report

Internal jugular vein tumor thrombus due to parapharyngeal extraosseous plasmacytoma

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ARTICLE INFO

Article history:

Received 4 March 2019

Revised 9 April 2019

Accepted 20 April 2019

Available online 14 May 2019

Keywords:

Plasmacytoma

Jugular vein

Tumoral thrombus

ABSTRACT

Tumor thrombosis of the internal jugular vein (IJV) represents an uncommon event, usually in the setting of underlying thyroid neoplasms. Extraosseous plasmacytoma (EMP) with tumor thrombosis of the IJV has not yet been reported in the literature. We present a unique case of a plasmacytoma in the left parapharyngeal space with direct extension to the left IJV, documented with contrast enhanced computed tomography and US Doppler. Presence of avid thrombus enhancement allowed differentiation between tumoral extension and thrombotic changes.

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Introduction

Plasmacytomas are rare malignant plasma cell tumours that arise without evidence of bone marrow plasmocytosis or systemic disease [1]. They are classified in 2 types: plasmacytoma of the bone and soft tissue plasmacytoma (extraosseous).

Solitary extraosseous plasmacytoma diagnosis requires local biopsy with proven monoclonal plasma cell histology, normal bone marrow aspiration and biopsy, and no abnormalities

attributable to plasma cell dyscrasia (anaemia, hypercalcemia, nor kidney disorders) [1].

If radiological skeletal survey is negative, whole-body MRI or ¹⁸F-FDG PET/CT scan is recommended to confirm a suspected diagnosis of solitary plasmacytoma [2–4]. CT and/or MRI are also recommended to evaluate the local extent of the lesion [4,5].

Median age of presentation is 55 years old with a 2:1 male preference [1,5–8]. Clinical presentation depends on the location and is usually related to compressive features and/or bleeding [9].

Abbreviations: CT, computed tomography; IJV, internal jugular vein; MRI, magnetic resonance imaging; PET/CT, positron emission tomography/computed tomography.

Acknowledgments: Special thanks to Isabel Candelária who provided language help and writing assistance.

Funding: This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

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<https://doi.org/10.1016/j.radcr.2019.04.012>

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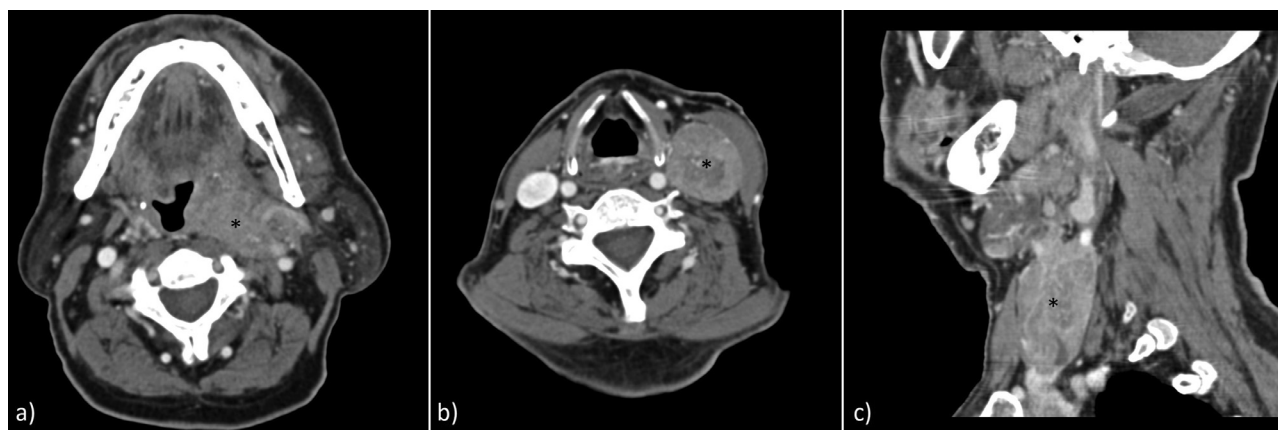


Fig. 1 – Contrast enhanced computed Tomography (CECT) showing a left parapharyngeal space centred mass (asterisk), compressing and displacing the left oropharynx wall (a). The mass extends laterally to the left IJV, expanding the vessel's lumen and demonstrating heterogeneous enhancement (b and c).

In 80%-90% of the cases, these tumours occur in the head and neck region, usually affecting the submucosa of upper respiratory tract (nasal cavity and paranasal sinuses mainly) [1,5–8].

The parapharyngeal space represents an uncommon location for these tumours [10] and no intravascular extension related to plasmacytoma has yet been described. Tumour thrombus of the jugular veins is mainly associated with thyroid malignancies [11,12].

Case report

A 68-year-old man was referred to our hospital due to a slow growing left cervical lump. An ultrasound was performed that suggested underlying adenopathy. Direct pharyngeal observation revealed a parapharyngeal left lump, without a mucosal lesion. Additional CECT revealed a massive deep cervical mass centred in the left parapharyngeal space, extending from the nasopharynx to submandibular level. The mass extended laterally to the internal jugular vein (IJV), with direct invasion of the vessel. An extensive intraluminal enhancing mass compatible with tumoral thrombus was revealed (Fig. 1).

Ultrasound guided biopsy of the cervical mass was requested, and during the procedure additional color Doppler exam performed. The evaluation confirmed left IJV thrombosis due to an intraluminal vascularized mass (Fig. 2).

The biopsy of the cervical mass confirmed the infiltration by clonal plasma cells. A monoclonal IgG kappa band protein was detected in blood. But no other abnormal laboratory findings were noted, in particular anaemia, hypercalcemia nor renal dysfunction. Additional skeletal survey did not suggest bone lesions. Bone marrow aspirate and biopsy did not demonstrate abnormal plasmacytosis. This set of findings, allowed to established the plasmacytoma diagnosis.

In a multidisciplinary meeting systemic treatment was proposed: bortezomib-dexamethasone and ciclofosfamide-

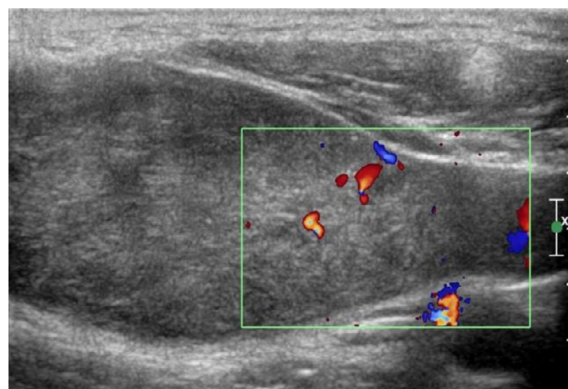


Fig. 2 – Ultrasound color Doppler showing the enhancing mass within of the internal jugular vein lumen.

vincristine-doxorubicin-prednisone, due to the extension of the lesion, but with no response.

Radiotherapy was then performed, with no significant tumour volume reduction, still not achieving partial response. Patient is actually under palliative treatment with good clinical status.

Discussion

Tumour thrombus in the IJV from a thyroid cancer was first documented in 1991 [13] but other head and neck tumours have been reported to invade or grow within the great vessels, such as the paragangliomas [14]. In our knowledge, plasmacytomas associated with venous tumoral thrombus has not yet been reported.

Thrombus associated with malignancy may result from either tumour vascular compression leading to stasis, or direct extension of the primary tumour. This distinction is essential

to the appropriate planning of surgical resection or radiation target volume delineation.

Differential diagnosis is based on the enhancement of the thrombus in post contrast imaging studies (CT or MRI), but may also be demonstrated in color Doppler ultrasound if the vessel location is accessible, such as the jugular vein, due to its superficial location.

Tumour thrombosis due to direct tumoral invasion shows enhancement after contrast, similar to the primary lesion. Thrombus due to coagulation has no post contrast enhancement.

Regarding plasmacytoma itself, no specific imaging features have been described, but lesions usually appear as a soft tissue mass, showing homogeneous enhancement with variable intensity degree [3].

Conclusion

IJV tumoral thrombus due to direct intraluminal vessel extension is a rare event, usually associated with thyroid malignancies, but may occur in other histologic neoplasms, including plasma cell neoplasms.

Its differential diagnosis from a thrombotic event is crucial to adequate treatment planning, specially if radiotherapy is a therapeutic choice.

Post contrast imaging features are diagnostic, revealing thrombus enhancement, but color Doppler ultrasound may also be performed in superficial located vessels.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2019.04.012](https://doi.org/10.1016/j.radcr.2019.04.012).

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