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Case report

Polymyalgia rheumatica and vagal paraganglioma

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ABSTRACT

Introduction: Vagal paraganglioma are rare tumors that are mostly asymptomatic. We report a case of vagal paraganglioma associated with paraneoplastic polymyalgia rheumatica and review the literature on benign paragangliomas of the head and neck associated with paraneoplastic syndrome.
Case report: A 53-year-old man presented with atypical polymyalgia rheumatica. MRI revealed a tumor that was then surgically excised. Histological examination confirmed the diagnosis of benign vagal paraganglioma. Rapid, complete and permanent resolution of all rheumatological symptoms were observed postoperatively, confirming the diagnosis of paraneoplastic polymyalgia rheumatica.
Conclusion: Paraganglioma of the neck associated with paraneoplastic syndrome remains exceptional. A predisposing gene mutation must be systematically investigated. Long-term surveillance must be ensured due to the risk of local recurrence, second tumors or metastasis.

1. Introduction

Paragangliomas are endocrine tumors arising from the neural crest: 0.3% are localized in the head and neck [1]. Carotid paragangliomas are the most common sites in the neck, followed by tympanojugular sites. Vagal paragangliomas are rare: only about 200 cases have been reported [1]. Paragangliomas of the neck are usually an incidental finding, as they remain asymptomatic or they may present as a neck mass or cranial nerve lesion [2].

We report a case of vagal paraganglioma diagnosed in a context of polymyalgia rheumatica and review the literature on paraneoplastic syndromes (PNS) in the context of benign paragangliomas of the neck.

2. Case report

A 53-year-old male active smoker with a history of intervertebral disc disease consulted with symptoms suggestive of atypical polymyalgia rheumatica. Laboratory tests revealed moderate and fluctuating inflammatory syndrome. PET-CT scan showed a suspicious left retromandibular hypermetabolic lesion (Fig. 1). After MRI assessment, the lesion was resected via a neck incision (Fig. 2).

Histological examination demonstrated a tumor with nodular architecture associated with abundant, richly vascular collagen stroma. The large tumor cells present abundant clear cytoplasm with a rounded nucleus and no nucleolus. Tumor cells expressed chomogranin and synaptophysin and stromal cells were labelled with anti-PS100 antibody. Finally, the Ki67 proliferation index was less than 1% (Fig. 3). Review of the slides by a REFCOR pathologist confirmed the diagnosis of paraganglioma with no signs of malignancy.

Plasma and urinary catecholamine assays were in favour of a non-secreting tumor and genetic analysis revealed the presence of a constitutional heterozygous c.43C > T(p.Arg15*) mutation of SDHC.

Complete resolution of the rheumatological symptoms and inflammatory syndrome were observed immediately after surgery. In view of completion resolution of rheumatological features after surgery, and absence of recurrence after three years, a diagnosis of vagal paraganglioma presenting in the form of PNS was adopted. Follow-up MRI at one year confirmed the absence of tumor recurrence and CT scan of the neck, chest, abdomen and pelvis at two-year follow-up demonstrated the absence of metastases.

3. Discussion

In this clinical case, the association between PNS (polymyalgia rheumatica) and benign vagal paraganglioma was based on clinical and histological data, resolution of the symptoms after surgery and the absence of distant metastases with a follow-up of 3 years.

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Table 1
Paragangliomas of the neck presenting as paraneoplastic syndrome.

	Authors	Site	Gender	Age	Paraneoplastic syndrome	Treatment	Outcome	Follow-up (years)
Benign paraganglioma	Tian et al.	Hypopharyngeal	Female	49	Neurological (encephalomyelitis)	Symptomatic: corticosteroid therapy Aetiological: surgical	Remission	Unknown
Malignant paraganglioma	Spraul et al.	Brain metastasis primary site?	Female	52	Exophthalmos	Palliative	Death in 18 months	2 years

Table 2
Main paraneoplastic syndromes, classified by organ or system, clinical and laboratory signs and underlying tumors.

Organ or system	Paraneoplastic syndrome	Symptoms	Laboratory parameters	Underlying cancers
Rheumatological	Polymyalgia rheumatica	Atypical: asymmetrical, steroid-refractory	ESR > 100/severe anaemia/proteinuria	Haematological malignancies Kidney/prostate/breast/colon/lung
	Rheumatoid arthritis	Rapid course, asymmetrical	Absence of rheumatoid factor	Haematological malignancies Lung/colon/breast
	Raynaud's syndrome	Asymmetrical course/thumb involvement/gangrene	Thrombocytopenia/ANCA/ANA	Haematological malignancies Liver/ovary/testis/kidney Melanoma
Endocrine	SIADH	Headache/asthenia/memory disorders Coma/respiratory collapse	Hyponatraemia/sodium excretion/urine osmolality	Small cell lung cancer
	Hypercalcaemia: PTHrp secretion bone resorption	Nausea/vomiting Drowsiness/coma	Serum calcium > 3.5 mmol/L ARF	Breast Multiple myeloma/lymphomas Bone metastases
	Cushing syndrome	HT Muscle weakness/oedema	Blood and urinary cortisol	Lung cancers
Neurological	Encephalitis/cerebellar degeneration/Lambert-Eaton syndrome/myasthenia gravis	Memory, cognitive disorders Ataxia Cranial nerve lesions	Onconeuronal antibodies	Lung cancers Lymphomas/multiple myeloma
Dermatological	Acanthosis nigricans	Cutaneous hyperpigmentation		Gastric adenocarcinoma
	Dermatomyositis	Erythematous rash Gottron papules	CPK/EMG Muscle biopsy	Breast/ovary/lung/prostate
	Leukocytoclastic vasculitis	Purpura/pain/pruritus		Haematological malignancies Lung/GI/urinary tract cancers
	Sweet's syndrome	Erythematous plaques, papules, nodules: face/trunk/extremities		CML/haematological malignancies Breast/urinary tract and genital/GI
Haematological	Eosinophilia	Asymptomatic ± dyspnoea		Haematological malignancies Lung/GI/genital
	Granulocytosis		WBC: 12 to 50×10^9/L	Lung/brain GI/kidney/genital
	Thrombocytosis		Platelets > 400,000/L	
	Pure red cell aplasia			Thymoma

ANCA: anti-neutrophil cytoplasmic antibody; ANA: antinuclear antibody; SIADH: syndrome of inappropriate antidiuretic hormone secretion; PTH: parathormone; ARF: acute renal failure; HT: hypertension; CPK: creatinine phosphokinase; EMG: electromyogram; GI: gastrointestinal; CML: chronic myeloid leukaemia; WBC: white blood cell.

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