Retroperitoneal ganglioneuroma causing chronic lower back and leg pain in an 80-year-old man: A case report

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ABSTRACT

Introduction: and importance: Retroperitoneal ganglioneuromas that cause lower back and leg pain are extremely rare and are often misdiagnosed. Surgical resection has excellent prognosis in long-term survival.

Case presentation: We present an 80-year-old man with two-year worsening left lower back and leg pain. He was treated as presumed lumbar spine spondylosis with several courses of physical therapy together with medical treatment. An abdomen CT scan disclosed a tumour in the left retrorenal space. The tumour was resected and the histopathologic examination suggested a completely excised retroperitoneal ganglioneuroma. During one-year follow-up the patient is free of pain without any local recurrence.

Clinical discussion: Retroperitoneal ganglioneuromas are rare benign tumors that originate from neural crest-derived cells of the paravertebral sympathetic plexus and sometimes from the adrenal medulla. They are usually asymptomatic and discovered on routine clinical examination or on autopsy. Occasionally they may show symptoms due to local pressure effect or rarely they are hormonally active and present with adrenergic symptoms. Complete resection of the tumor is important in order establish the final diagnosis and alleviate symptoms from pressure effects.

Conclusion: This case highlights the need for great vigilance among physicians in order to consider any possible retroperitoneal pathology when indicated in the differential diagnosis of lower back and leg pain, before establishing other more common diagnosis, especially in the older population.

1. Introduction

Ganglioneuromas are rare benign well-differentiated neoplasms that are composed by Schwann cells on nerve fibres and mature sympathetic ganglion cells [1]. They usually originate from the paravertebral sympathetic plexus extending from the skull to the neck, posterior mediastinum and retroperitoneum. Approximately 15%–30% originate from the adrenal medulla [2,3]. Other unusual areas of occurrence include the gastrointestinal tract, bones, supraclavicular area and pelvis [4]. We describe an 80-year-old man with left lower back and leg pain for the last two years, due to the presence of a retroperitoneal ganglioneuroma, who was treated as presumed lumbar spine spondylosis. This case highlights the high index of suspicion required in order to consider any possible retroperitoneal pathology in the differential diagnosis of lower back and leg pain, before establishing other more common diagnosis, especially in the older population. This case report has been reported in line with the SCARE Criteria [5].

2. Case presentation

An 80-year-old Cypriot man, smoker (80 pack-yr) presented to our clinic with his daughter complaining for gradually worsening dull ache pain to his left lower back for the last two years. The pain occurred independently of any physical activities and radiated to his left anterior
thigh with concomitant numbness in this area. Weight loss of approximately 4 kg (kg) during the last eight months was also reported. He was treated during this period as presumed lumbar spine spondylosis, based on the results of plain radiography of the lumbar spine, suggesting moderate degenerative changes. He experienced several courses of physical therapy by a chiropractor together with medical treatment. Non-steroidal anti-inflammatory drugs, muscle relaxants, opioids, antiepileptics, antidepressants and epidural corticosteroid injections were administered without any meaningful improvement of his symptoms. His past medical history was significant for chronic obstructive pulmonary disease. His current medications were pregabalin 150 mg every 12 hours, duloxetine 60 mg daily, paracetamol/codeine phosphate 500 mg/30 mg every 8 h and fluticasone furoate/vilanterol 92/22 μg daily. Neither past surgeries nor any family history of genetic disorders was reported. His mental health was excellent.

On physical examination the patient appeared to be in good condition having 13 breaths per minute, temperature of 36.5 °C, blood pressure of 120/70 mmHg and heart rate of 88 beats per minute. His body mass index (BMI) was 24 kg/m² and waist circumference 95 cm. Moderate hypoesthesia of the left anterior thigh and 4+/5 strength of left knee extension were found in the neurological examination. The rest of the clinical examination was unremarkable except of diminished bilateral breath sounds during chest auscultation. Mild normocytic normochromic anemia (hemoglobin 12.6 g/dl), white blood count 10.780/mm³ (neutrophils: 81%), erythrocyte sedimentation rate (ESR) 5 mm/hour and C-reactive protein 0.41 mg/dl (normal values < 0.5 mg/dl) were found. Other laboratory investigations were within normal limits including liver and renal function indices, electrolytes and thyroid profile.

The unrelenting pain at rest together with body weight loss during the last months raised concerns for other possible more serious causes. An abdomen computer tomography (CT) scan showed a hypodense lesion of 3.5 cm maximum diameter in the left retrorenal space (red arrows) (Fig. 1). A chest CT scan disclosed low attenuation areas as well as vascular thinning suggesting pulmonary emphysema. Surgical excision of the tumour was made by a consultant surgeon specialised in gastrointestinal oncology and a consultant urologist. During the surgery a mass with well-defined borders in close proximity to the left lateral body of the spine [extending from the bottom of lumbar dermatome 1 (L1) to the top of L3 level], being firmly attached to the lumbar fascia, was identified. It originated posterior to the left renal vessels and anterior to the upper limit of the psoas muscle compressing the lumbar vessels. Gross examination revealed a 32 g well circumscribed encapsulated tumour measuring 6 × 4 × 3.5 cm (Fig. 2). On sectioning the tumor was white to tan-yellow and firm. Histopathological examination showed a hypercellular neoplasm, characterized by the presence of mature ganglion cells individually distributed in Schwannian fibrous stroma without any signs of malignancy (Fig. 3). Immunohistochemically, the neoplastic ganglion cells were strongly positive for synaptophysin, S-100 and weakly positive for chromogranin and glial fibrillary acidic protein (GFAP). The Schwannian stroma was diffusely S100 and GFAP positive. A completely excised retroperitoneal ganglioneuroma was concluded. The patient experienced an uneventful postoperative period. Patient adherence to all medical advice given, after discharged with good recovery.
from the hospital, was excellent. During one-year follow-up the patient is free of pain, without receiving any relevant medications, something that was beyond his expectations as he described. No local recurrence was found.

3. Discussion

Retroperitoneal ganglioneuromas that cause lower back and leg pain in this age group are extremely rare. The first case report was published by Anderson et al. in 1953 [6]. Unfortunately, the possibility of a retroperitoneal tumour as a cause of these symptoms is rarely considered in middle-age or elderly population. It is almost always regarded as an epidemic symptom of lumbar spondylosis especially if degenerative lesions are found in radiology [6]. Hence the diagnosis is often delayed like our patient. The differential diagnosis includes other systemic disorders, mainly primary metastatic neoplasms (including multiple myeloma) and inflammatory spondyloarthropathy, as well as gastrointestinal disorders, genitourinary disorders (including nephrolithiasis), abdominal aortic aneurysm and hip pathology. The patient’s current medical history, his clinical examination, as well as all laboratory and radiology studies excluded all other possible causes. Chronic lower back pain radiating into the anterior thigh, described independently of activities and poorly responding to physiotherapy or any possible medical treatment, could be explained due to nerve compression from the tumour and irritation of certain branches of the lumbar plexus in the retroperitoneal space [7,8].

The majority of ganglioneuromas are non-secreting tumors. They are usually asymptomatic and discovered on routine clinical examination or on autopsy. Occasionally they may show symptoms due to local pressure effect or rarely they are hormonally active [2,4,9]. They can occasionally release catecholamines and cause adrenergic clinical symptoms (sweating, diaphoresis, tachycardia, paroxysmal hypertension, headache) similar to those of mature chromaffin secreting tumors [9,10]. Ganglioneuromas can rarely cause weight loss like our patient [11]. Isolated adrenal ganglioneuromas have been also associated with multiple endocrine neoplasia 2A (MEN2A) and MEN2B, without evidence of pheochromocytomas [12]. Local extra-adrenal extension or malignant transformation of adrenal ganglioneuroma can be rarely observed [13].

Treatment is complete surgical resection and has an excellent prognosis in long-term survival. Residual disease after incomplete resection can remain stable for years without malignant transformation. Less aggressive surgical approach, in order to preserve the adjacent structures and organs, when is appropriate has been suggested [14-16].

4. Conclusions

This case highlights the need for great vigilance among physicians in order to consider any possible retroperitoneal pathology when indicated in the differential diagnosis of lower back and leg pain, before establishing other more common diagnosis, especially in the older population. Complete resection of retroperitoneal ganglioneuromas alleviates any possible symptoms and has an excellent prognosis in long-term survival.

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All authors; Georgios S. Papaetis, Maria A. Tsiskari, Pavlos G. Constantinou, Christos P. Georgiadis and Antonis P. Antoniou had involved in this case report design, data collection and manuscript preparation. They reviewed the final version and literature of the project and all agreed for publication.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Declaration of competing interest

The authors declare that they have no competing interests.

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