Bone Reports 22 (2024) 101779

Contents lists available at ScienceDirect

Bone Reports

journal homepage: www.elsevier.com/locate/bonr

Case Report

Pseudotumoral calcinosis of the elbow with ulnar and radial nerve deficits

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ARTICLE INFO

Keywords:
Pseudotumoral calcinosis
Hemodialysis
Elbow surgery
Nerve compression

ABSTRACT

We report here a rare case of an acute peripheral nerve compression by pseudotumoral calcinosis (PCT) at the right elbow in a patient with severe tertiary hyperaparathyroidism. This complication required urgent multidisciplinary management. Surgical decompression by PCT resection enabled rapid motor and sensory recovery.

1. Introduction

Pseudotumoral calcinosis (PTC) is a rare complication of chronic hemodialysis characterized by calcium phosphate deposits in the periarticular tissues, of poorly known pathophysiology, which seems to be enhanced by severe hyperparathyroidism (Kabbali and Sqalli, 2017). We report here the case of a 40-year-old patient with PTC of right elbow requiring surgical management due to acute peripheral neurological complications.

2. Case

A 40-year-old man was admitted at the Rheumatology Department for swelling of the right elbow and left shoulder, causing pain and impaired function. His past-medical history was characterized by an end-stage renal disease on focal segmental hyalinosis requiring hemodialysis since 2005 and a kidney transplant complicated by rejection in 2008. Clinical examination revealed a voluminous mass of the right elbow and left shoulder, resulting in active and passive limitations of joints mobility, progressing for 18 months. There was no initial neurological deficit. Beside the end-stage renal failure (CKD EPI 6 ml/min/1.73m^2), biological assessment revealed moderate chronic hypercalcemia (2.64 mmol/l, normal range 2.15–2.55 mmol/l), high hyperphosphatemia (2.77 mmol/l, normal range 0.84–1.45 mmol/l), normal 25(OH)vit D level (56 ng/l, normal >30 ng/ml), increased PTH (415 ng/l, normal range 18.4–80.1 ng/l) and a chronic biological inflammatory syndrome (CRP 40 mg/l, normal <5 mg/l). Radiological right elbow assessment showed voluminous calcifications with no joint destruction (Fig. 1). Ultra-sound and anterior dual-isotope ($^{(123)}$I/$^{(99m)}$Tc-Sestamibi) static planar imaging followed by hybrid SPECT/CT revealed 4 parathyroid lesions over 1 cm, vascularized, including at least 2 autonomous glands. A subtotal seven-eighths parathyroidectomy was planned. In the meantime, a motor deficit in the posterior interosseous nerve territory occurred, with a rapid deficit evolution from 4 to 1/5 in 10 days for finger extension and a slight deficit in wrist extension. An associated sensory deficit with major pain in the radial territory was reported. Ulnar sensitive sign appeared over the last few days. Nerve compression at the elbow was confirmed by electromyography which concluded that there was a motor conduction block at the elbow on the left ulnar with slight motor denervation of the first dorsal interosseous tract. In emergency, the surgical team performed the surgical excision of 2 pseudotumor masses of 12 × 7 cm and 10 × 10 cm developed from the anterior joint capsule of the elbow (Fig. 2). A neurolysis of the superficial and deep branches of the radial nerve was associated. The extension of the tumors was mainly lateral and posterior. Tumors were fairly well-circumscribed with several muscular insertions and capsular continuity. The anatomopathological analysis confirmed the diagnosis of a PTC, without malignancy criteria (Fig. 2). After nerve decompression, we observed a rapid and complete clinical improvement in sensory and motor signs. Few days after elbow surgery, the parathyroidectomy was

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https://doi.org/10.1016/j.bonr.2024.101779
Received 15 February 2024; Received in revised form 10 June 2024; Accepted 12 June 2024
Available online 13 June 2024

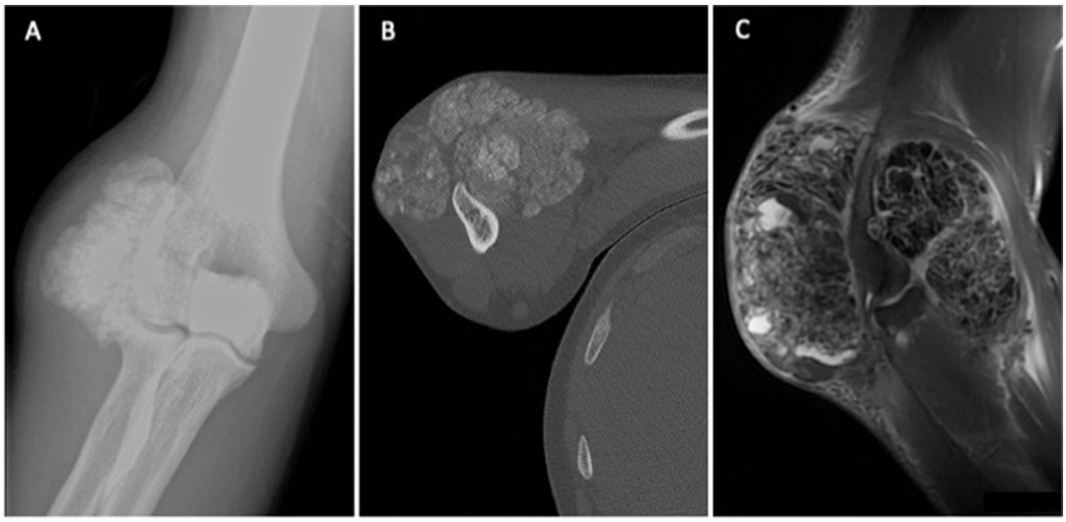

Fig. 1. A. En-face view X-rays of the right elbow with periarticular calcifications. B. CT-scan of the right elbow objectived two calcified masses (anterior and lateral) C. MRI of the right elbow shows extrinsic compression of the deep branch of the radial nerve against the humerus by the anterior masse.

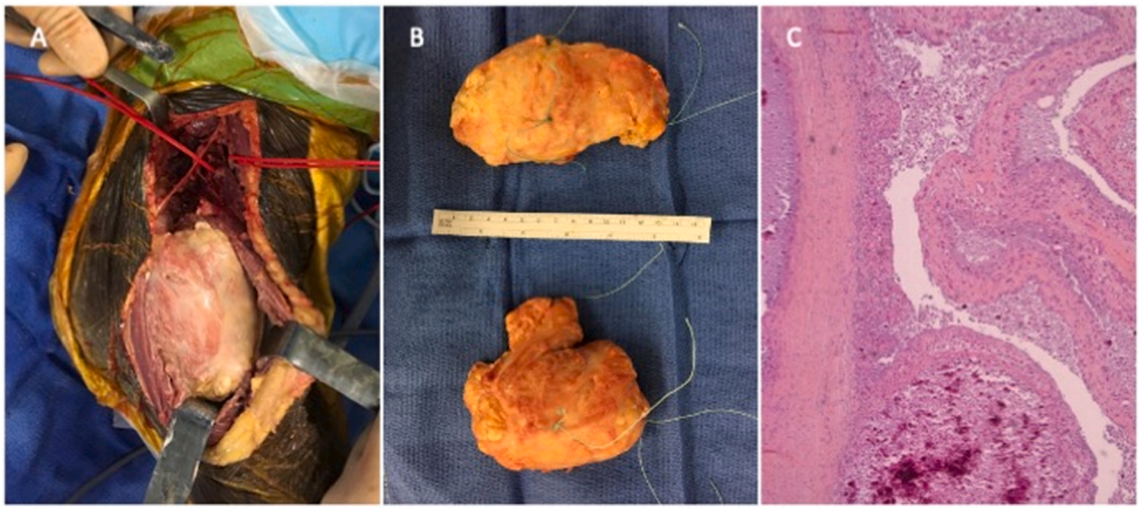

Fig. 2. A. Intra operative image of the elbow. B. Morphology of the PTC after surgical resection. C. Anatomopathological analysis: calcified central material surrounded by a macrophagic reaction with many plurinuclear giant cells associated with a few mononuclear inflammatory elements.

performed. Anatomopathological analysis confirmed the tertiary hyperparathyroidism.

3. Discussion

The originality of this case was the presence of 2 PTC that generated rapid nerves compression. To our knowledge only few cases were described before: one in a patient suffering from scleroderma with PTC of the wrist compressing radial nerve (Driouach et al., 2020), and 2 cases of uremic tumoral calcinosis responsible of carpal tunnel syndromes (Asami et al., 1998; Cofan et al., 2002). In our case, we must note that compliance with treatment was poor for this patient who wanted to continue to work-full time: dialysis skipping, incorrect phosphate chelators intake (sucroferric oxyhydroxide and sevelamer carbonate), with episodes of significant increases in phosphate levels up to 3.3 mmol/l, identified as the main cause of evolutive PTC.

In a pathophysiological point of view, in long standing CKD, hyperphosphatemia causes direct stimulation on parathyroid cells causing nodular hyperplasia and PTH hypersecretion. Increase FGF23 in turn usually reduces the PTH secretion, but FGF23 receptor expression on parathyroid gland can be decreased, causing uninhibited PTH secretion. A decreased formation of 1,25 (OH) vitamin-D by decreased 1α hydroxylase activity in kidney, lead to secondary hyperparathyroidism characterized by hypocalcemia and increased PTH secretion. Moreover, decreased expression of vitamin-D receptor on parathyroid tissue results in increased expression of PTH gene. Calcium sensing receptor set point is also altered. Thus, longstanding CKD and secondary hyperparathyroidism leads to an autonomous secretion of PTH and a persisting hypercalcemia, which may not be reversed after renal transplantation, characterizing tertiary hyperparathyroidism (Jamal and Miller, 2013).

Therefore, we assume this diagnosis deserves to be reported. Although soft tissue calcification is common in secondary/tertiary hyperparathyroidism due to high calcium-phosphate product, compressive PCT is a very rare condition. This case underlines the importance of the management of hemodialysis patients presenting tertiary hyperparathyroidism and PTC: place of surgical management of such a potentially damaging tumor and optimization of the treatment of tertiary hyperparathyroidism.

Ethical form

The authors declare that they have obtained written and signed consent to publish the case report from the patient.

CRediT authorship contribution statement

Marie-Charlotte Trojani: Writing – original draft, Formal analysis, Data curation. **Marc-Olivier Gauci:** Supervision, Data curation. **Caroline Cointat:** Data curation. **Laura Cabane:** Data curation. **Véronique Breuil:** Writing – review & editing, Conceptualization.

Declaration of competing interest

There are none.

Data availability

Data will be made available on request.

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