CASE REPORT

BONE AMYLOIDOMA IN A DIABETIC PATIENT WITH MORBID OBESITY

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SUMMARY - Bone localisations of amyloidosis are rare, usually diffuse and associated with myeloma. We report the case of a patient with massive obesity complicated by diabetes, hypertension, sleep apnea and liver steatosis, who complained of rapidly worsening bilateral polyradiculalgia of the lower limbs. After sufficient weight loss made nuclear magnetic resonance imaging feasible, a spinal tumour was visualised on the 5th lumbar vertebra, extending to soft tissues. Total excision was performed, and pathological studies revealed an amyloid bone tumour with no evidence of myeloma.

Key-words: amyloidoma, diabetes, neuropathy, morbid obesity.

RÉSUMÉ - Amylose osseuse chez un patient diabétique avec obésité morbide.
Les localisations osseuses de l’amylose sont rares, le plus souvent diffuses et associées au myélome. Nous rapportons le cas d’un patient présentant une obésité massive compliquée d’un diabète, d’une hypertension artérielle, d’un syndrome d’apnée du sommeil et d’une stéatose hépatique, se plaignant de polyradiculalgies bilatérales des membres inférieurs s’aggravant rapidement. Après un amaigrissement suffisant l’imagerie par résonance magnétique devenue possible a montré une tumeur osseuse lombo-sacrée en sablier, centrée sur la cinquième vertèbre lombaire et s’étendant aux parties molles. Après exérèse totale l’examen anatomopathologique a permis le diagnostic de pseudo-tumeur amyloïde localisée à l’os, chez un patient ne présentant aucun signe de myélome.
Mots-clés : pseudo-tumeur amyloïde, diabète, neuropathie, obésité morbide.

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Amyloidosis is defined as the extracellular deposit of a fibrous protein which constitutes the amyloid substance in one or several tissues [1]. Various types have been described: primary amyloidosis and amyloidosis associated with multiple myeloma (both of the AL subtype); amyloidosis secondary to infectious or chronic inflammatory diseases (AA subtype); and isolated amyloidosis in an organ without systemic disease. Amyloidosis in bone is infrequent, and usually diffuse in patients with multiple myeloma and subjects undergoing chronic dialysis [2]. We report the extremely rare case of a Type 2 diabetic patient with morbid obesity and polyradiculalgia of the lower limbs, who presented pseudotumoural amyloidosis in bone without any clear etiology.

Case report

A 45-year-old man was referred to our department in January 1993 for morbid obesity. His medical history included rheumatic fever in childhood and an abdominal lipectomy six years before, complicated by eventration. He weighed 190 kg and was 1 m 74 cm tall (body mass index: 62.8 kg/m²). Obesity was of android type (waist-to-hip ratio: 1.2), mainly complicated by Type 2 diabetes which was diagnosed in 1991 and poorly controlled by glibenclamide 15 mg and metformin 1,700 mg in the absence of a suitable diet. His hypertension was poorly controlled by calcium-channel blockers, and he suffered from joint pain in the lower limbs. He also complained of recent sexual impotence. Neurological examination was normal, and neurophysiological investigation showed early signs of right carpal tunnel syndrome and sensorial neuropathy of the lower limbs. Biological investigations revealed a moderate inflammatory syndrome persisted. The patient described paresthesias and polyradiculalgias in the lower limbs. A CT-scan was scheduled but could not be performed because of the patient's corpulence. In February 1996, body weight was 148 kg, and the symptoms persisted despite antalgic and anti-inflammatory treatment. Neurological examination showed hypoesthesia of the soles of the feet, whereas tendon reflexes and superficial and proprioceptive sensitivity were normal. Electrophysiology showed a sensorimotor axonolytic neuropathy. Blood glucose and liver tests were normal, as were inflammatory parameters.

Bilateral lumbar sciatia became constant, inducing sleeplessness. In September 1996, radiologic assessment was finally feasible after considerable weight loss. CT-scan and MRI of the back revealed a bone lesion centred on the posterior part of the 5th lumbar vertebra, with extension into soft tissues and the lumbar canal between the 4th lumbar vertebra and the sacrum as well as destruction of the corresponding posterior joint apophysis. A percutaneous biopsy suggested benign dermoid cyst. In October 1996, surgery was performed with great difficulty because of the patient's morphology. Macroscopic examination confirmed the radiologic images but ruled out the hypothesis of a dermoid cyst. The tumour was totally removed. Recovery after surgery was uneventful, and the patient became fairly self-sufficient after experiencing considerable symptom relief.

Histological examination of the lesion revealed large round sicks, with a stratified appearance, occasionally surrounded by macrophages or giant cells. These deposits also infiltrated bone medullary cavities. Labelling with Congo red and T thioflavine revealed amyloid material. Cell aggregates (mainly plasma cells) were observed around some vessels and medullary cavities. Many of these cells were dystrophic, often with two or three nuclei. Immunohistochemical study showed marked positivity for the light kappa chain and a lack of stamping for heavy chains and the light lambda chain.

Biological examination showed a normal blood cell count and erythrocyte sedimentation rate. Plasma protein electrophoresis and immunoelectrophoresis were normal, as were plasma immunoglobulins and $\beta_2$-microglobulin. There was no Bence-Jones proteinuria. Bone-marrow puncture failed to reveal any medullary plasmacytosis. Pelvis and skull radiographs were normal.

Discussion

Although all organs and joints (in particular) may be affected by amyloidosis, focused bone localisations have been reported only rarely [2-4]. Bone amyloidosis is usually diffuse, producing various types of clinical and radiological images: osteoporosis, destruc-
tive lesions, pathological fractures, osteonecrosis, nodes and infiltration of soft tissues, cysts and subchondral erosions, and joint subdislocations with contracture and neuropathic osteoarthropathy [5]. Amyloid deposits in the skeleton are often associated with multiple myeloma related to diffuse amyloidosis with plasmacytic infiltration [2]. Amyloid bone tumour is very rare, corresponding to the accumulation of amyloid material in an amount sufficient to destroy bone and become visible on radiography, mainly in the form of destructive lesions, often with fractures [1]. All bones may be affected, especially long ones, the spine and the base of the skull [6, 7]. Symptoms are various and non-specific, ranging from pain to complete paraplegia [8]. Other diagnoses may be considered, mainly primary bone tumour, metastasis or metabolic diseases such as gout. Macroscopically, a solid homogeneous yellow or brown mass is apparent. Microscopic examination shows lifeless, vitreous eosinophilic material, sprinkled with giant multinuclear cells, histiocytes and plasma cells [9]. Several stains have been used to distinguish between amyloidosis and other kinds of proteins, but the most common one is the Congo red preparation which shows red-green birefringence in polarising microscopy. The plasma cells found in these amyloid tumours suggest a possible link with solitary myeloma [10].

Multiple myeloma is by far the main etiology for these localised lesions, but in a few cases large bone amyloid deposits have been found without plasma cell infiltration [1]. In some cases, no cause has been determined, despite Bence-Jones proteinuria or extra-bone localisations (heart, kidney, pancreas, nodes) [9]. Subjects under chronic dialysis are at great risk for amyloid tumours due to the deposit of B2-microglobulin in the skeleton, which usually causes large destructive lesions leading to fractures [11]. In our patient, apart from a moderate inflammatory syndrome, there was no evidence of myeloma. He never exhibited clinical or radiological symptoms suggestive of another bone disease, nor had significant proteinuria. Moreover, histological examination failed to show amyloid localisation in the liver.

This case report illustrates another effect of massive obesity. In addition to the classical metabolic, cardiovascular and rheumatological complications, the patient’s massive overweight condition made it impossible to use the most up-to-date imaging techniques, such as CT-scan or MRI. This may seem anec-

dotal, but the consequences can be crucial in life-threatening circumstances when these diagnostic means are essential. In our patient, if it had been possible to make the diagnosis earlier, many months of suffering and disability would have been avoided and surgery would have been safer and less traumatic. Moreover, the patient’s morphology, in addition to presenting a marked anaesthetic risk and obvious technical difficulties related to the impressive thickness of fat tissues, had a definite influence on the surgical procedure itself since arthrodesis could not be considered as the first indication. Arthrodesis is performed to avoid secondary spondylolisthesis, but the genupectoral position must be maintained for at least five hours, which greatly increases the risk of postoperative rhabdomyolysis.

Polyradiculalgias are a rare manifestation of diabetic neuropathy. This case report indicates the need to exclude causes of compression by making an appropriate assessment.

REFERENCES