CASE REPORT

Diagnosis of an Immunoglobulin D multiple myeloma with severe renal involvement a decade after a solitary plasmacytoma: A case report and literature review

Diagnostic d'un myélome multiple à immunoglobuline D Lambda avec une atteinte rénale sévère 10 ans après un plasmocytome solitaire : Un cas clinique et revue de la littérature

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Abstract

Introduction: Immunoglobulin D (IgD) myeloma is a rare subtype often described as aggressive with advanced disease at diagnosis. Primary renal involvement is seen in scarce cases.
Observation: This case features a 55-year-old man with IgD lambda myeloma presenting severe renal failure at diagnosis. Examination revealed a 10-year-old sternal plasmacytoma and multiple others in the ribs. Despite benefiting from traditional chemotherapy, he remained dependent on hemodialysis
Conclusion: Through this case, unique in the literature, we conclude that plasma cells secreting IgD can remain inactive for a long time in the form of a solitary plasmacytoma. However, in the event of medullary involvement, they can induce a myeloma with serious organic lesions.

Key words: bone; dialysis; neoplasm; plasma cell; uremia;

Résumé

Introduction: Le myélome à immunoglobuline D (IgD) est un sous-type rare souvent décrit comme agressif avec une maladie avancée au moment du diagnostic. Une atteinte rénale primaire est observée dans de rares cas.
Observation: Nous décrivons le cas d'un homme de 55 ans atteint d'un myélome à IgD-lambda avec une insuffisance rénale sévère au moment du diagnostic. A l'examen, il présentait une voussure sternale datant de 10 ans. Les investigations ont conclu à un plasmocytome sternal associé à plusieurs autres dans les côtes. Il a bénéficié d'une chimiothérapie et est resté dépendant de l'hémodialyse.
Conclusion: A travers ce cas, unique dans la littérature, nous concluant que les plasmocytes sécrétant des IgD peuvent rester longtemps inactifs sous la forme d'un plasmocytome solitaire. Cependant, en cas d'atteinte médullaire, ils peuvent induire un myélome avec de graves lésions organiques.

Mots clés: dialyse; néoplasme ; os; plasmocyte ; urémie

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Introduction

Immunoglobulin D (Ig D) multiple myeloma (MM) is a rare subtype, comprising less than 2% of all cases (1). It often presents an aggressive clinical course and a poorer prognosis compared to other MM variations (1, 2).
IgD MM shares clinical features with IgG and IgA myeloma but shows a higher prevalence of skeletal manifestations (3). Despite this, severe kidney damage is uncommon, occurring in only 15% to 20% of cases (4).
Solitary plasmacytoma is an unusual plasma cell disorder distinguished by the presence of a single bone lesion infiltrated by monoclonal plasma cells, without extensive bone marrow infiltration or damage to vital organs (5).
Our case study highlights an unusual instance of IgD MM with severe kidney impairment a decade after a dormant solitary sternal plasmacytoma was identified. This is the first case reported, to our knowledge, in the literature.

Case report

A 55-year-old man with no significant medical history was admitted to the hospital for investigation of severe kidney failure, which was found after experiencing vomiting and abdominal pain.
During the physical examination, the body mass index was 33 kg/m2, with systolic and diastolic blood pressure at 140 and 90 mmHg, respectively. The patient's hydration status was normal. Urinalysis showed trace amounts of leukocyte esterase, protein, and blood on dipstick. Notably, a hard, painless 7x4 cm oval swelling without signs of inflammation was observed adjacent to the sternal body, which the patient reported to have been developing for approximately ten years (Figure 1).

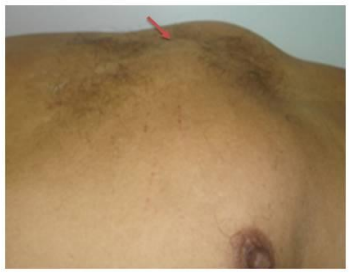

Figure1. Curvature originating from the sternum and evolving over 10 years

Serum laboratory study showed normochromic anemia and severe renal failure (Table 1).
Abdominal ultrasound revealed kidneys of normal size with regular cortical thickness, absence of hydronephrosis or urolithiasis, and no presence of masses.
Even though the test strip only showed small amounts of proteinuria, the 24-hour proteinuria test revealed a notable presence of protein in the urine (Table 1).
This observation related to normochromic anemia and normal sized kidneys was a sign of possible paraproteinemia-associated kidney disease. The subsequent analysis of serum proteins showed a single peak moving into the γ globulin region. Blood Ig levels indicated a rise in IgD levels accompanied by a reduction in other Ig levels. Further examinations involving serum immunofixation and serum-free light chain testing identified significantly elevated serum lambda-free light chains and a markedly high lambda/kappa ratio.

Table 1. Serum laboratory results

	Findings	Normal range
White blood cell count (k/uL)	5.8	4.0-10
Platelet count (k/uL)	328	150-450
Hemoglobin (g/dL)	6.0	13.5-18.0
Mean corpuscular volume (fL)	86	80-95
Glucose (mmol/l)	4.8	4.1-5.9
Creatinine (µmol/l)	1390	64-104
Blood urea nitrogen (mmol/l)	13.4	2.8-7.2
Bicarbonate (mmol/l)	20	17-24
Calcium (mmol/l)	2.02	2.20-2.65
Lipase (U/L)	30	11-82
Total protein (g/L)	80	66-83
Albumin (g/L)	38.8	36.0-48.0
γ globulin (g/L)	21	8-13.5
Immunoglobulin D (g/L)	2	0.05–0.40
Serum lambda-free light chains (mg/dl)	1120	0.33-1.94
Serum lambda/kappa ratio	702.00	0.26-1.65
Proteinuria (g/24hours)	3	<0.5

Severe renal failure with significantly elevated levels of free light chains indicates monoclonal gammopathy. To confirm this diagnosis, the patient underwent bone marrow and kidney biopsies. The bone marrow biopsies showed abnormal plasma cells, making up to 60% of myeloid cells in certain regions. The renal biopsy revealed seven normal glomeruli and renal tubules with protein material casts that varied from glassy to slightly granular. The tubular epithelium showed signs of weakening and flattening, indicating cast nephropathy.
The sternal mass puncture revealed a sample with mostly abnormal plasma cells. A sternal plasmacytoma was identified in the skeletal survey, along with several osteolytic lesions in the sternum and ribs (Figure 2).

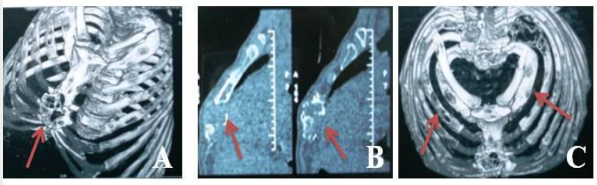

Figure 2. Chest computed tomography scan sequencies: Sternal plasmacytoma (**A and B**), multiple costal plasmacytomas (**C**)

The diagnosis of IgD MM stage III B of the International Staging System (6) with myeloma cast nephropathy and multiple plasmacytomas was confirmed.
The patient unfortunately could not benefit from a treatment based on Bortezomib and Thalidomide. After three blocks of dexamethasone 15 days apart and alkalinization of the urine, there was no improvement in renal function, leading to the need for hemodialysis. On a positive note, there was a 50% reduction in the size of the sternal mass.

The patient received then monthly boluses of melphalan (at a dose of 5 mg/m2) and prednisone (60 mg/d) 4 days in a row.
Six months later, he undergoes hemodialysis. Activity parameters did not return to normal. Radiotherapy was not recommended for plasmacytomas due to the lack of functional complaints.

Discussion

In this article, we reported a rare case of severe renal damage of myelomatous origin, occurring several years after bone damage.
MM is a type of aggressive cancer that involves the abnormal growth of plasma cells infiltrating the bone marrow. (7) Median age at diagnosis is 66 years (8). Malignant plasma cells can impact multiple organs, leading to various symptoms like bone pain, kidney failure, hypercalcemia, anemia, fractures, and signs of hyperviscosity. (9, 10). Occasionally, neoplastic plasma cells can exhibit a distinct growth pattern resulting in the formation of tumor masses known as extramedullary plasmacytomas (EMP) in 9% of cases. (11).
The majority of patients with MM (97%) produce a unique protein from cancerous plasma cells. (8). These cells can secrete heavy and light chains, light chains, or do not produce Immunoglobulins (8). Their frequencies are IgG (52%), IgA (21%), kappa or lambda light chain only (16%), IgD (2%), X (2%), IgM (0.5%) and negative (6.5%) (8).
IgD MM is a less common form of the disease that is more prevalent among the Asian population (12). It typically occurs at a younger age, mostly in males, with an average age of onset at 58 years. (12, 13).
Plasma cells that secrete IgD are formed due to genetic changes in the IgD area of B cells in the germinal center. (10). In IgD myeloma, a unique feature is the predominance of lambda light chains, which is present in 70% to 90% of cases. (10). The clinical aspects of IgD MM are comparable to those of other myeloma types (1). Weakness, pallor and bone pain are the earliest manifestations (10). The risk of kidney injury in MM IgD is increased and patients with renal failure tend to be younger (10). Renal failure is reported in 20 to 40% of patients at the time of diagnosis according to literature sources (PubMed) (13) while primary renal injury and severe renal failure are observed in only 15 to 20% of cases (4,14, 15).
The process of renal damage mechanisms of renal injury may be due to light chain nephropathy or direct toxicity induced by intracellular crystals (10,14). In newly diagnosed IgD MM patients, renal failure at diagnosis is associated with a poorer prognosis (13). Extramedullary participation in MM IgD is observed in approximately 19% to 63% of patients. Typical locations include the chest wall, airway, gastrointestinal tract, skin, lymph nodes, paraspinal area, and occasionally the testicles (10, 15). EMP may appear at diagnosis or develop later in the course of the disease (10). It can manifest as an extradural tumor or nerve root compression, and research indicates that patients with EMP experience decreased progression-free survival. (1).
Our patient, hailing from North Africa, was diagnosed with IgD lambda MM at 55 years old. Initially, he experienced vague symptoms such as abdominal pain and vomiting. Notably, he exhibited significant renal involvement and rare severe renal failure, uncommon in IgD myeloma cases. Particularly striking in this case was the discovery of a 10-year-old sternal mass, initially disregarded, eventually identified as a plasmacytoma. This suggests a decade-long presence of a dormant sternal plasmacytoma that later progressed into MM.
According to the literature, patients with isolated plasmacytoma and little bone marrow involvement have a higher risk of progression to MM than when no plasmacytosis is detected in the marrow (5). This was first demonstrated in a retrospective review of 127 patients at the Mayo Clinic (16). In their study, which included 50 patients, Hill et al. (17) concluded that progression occurred in 72% of patients with detectable latent bone marrow disease, compared with 12% in patients without detectable bone marrow disease. The median time to onset of MM was 21 months, with a 5-year probability of 51% in the series of Knobel et al. (18).
Our patient most likely had a solitary plasma cell for a long time without bone marrow involvement. This would explain the very late evolution towards an MM.
Historically, IgD MM has been thought to be associated with a poor prognosis (12, 19). Compared with other subtypes, the median survival of patients with IgD type received traditional chemotherapy was generally less than two years (20). However, some recent studies have suggested comparable survival rates with other subtypes thanks to autologous hematopoietic stem cell transplantation and novel therapies such as proteasome inhibitors and immunomodulatory agents (12). Unfortunately, our patient could not benefit from these therapies.
The case report describes a single patient, which limits the generalizability of our findings. Additional case reports may validate the conclusion drawn from this case.

Conclusion

From this observation, we conclude that IgD-secreting plasma cells, which are known to induce highly aggressive MM, can remain quiescent for a long time in the form of a solitary plasmacytoma. However, in case of bone marrow involvement, they can induce MM with severe organic damage.

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