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Case Report

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"Patient with Retroperitoneal primary Plasmocitoma and progression to Intracranial Plasmocitoma in third ventricle, Case Report"

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Introduction

The first neoplastic proliferation of plasma cells was described by Dalrymmple and Bence-Jones in 1846. They described a disseminated neoplastic proliferation of plasma cells characterized by marked proteinuria and bone pain. Rustizky in 1873 coined the term multiple myeloma (MM) for this disease. MM is a type of hematologic neoplasm that usually shows diffuse or multiple bone involvement and is characterized by the proliferation of a single plasma cell clone, but with varying degrees of immaturity, including atypical forms. Although its location is common in the bone marrow, multiple myeloma has been described as a tumor in a unique location of the skeleton, in these cases it is called solitary plasmacytoma, and in areas outside the skeleton, in which cases it is called plasmacytoma. extramedullary The primary extramedullary plasmacytomas are infrequent, almost always appear solitary and without systemic affectations. Its most common localization sites are: the upper respiratory tract, digestive system, genitourinary system, lung, lymph nodes and skin, the cranial brain location is extremely rare (0.7%).Solitary plasmacytomas originate in any organ due to the universal distribution of plasma cells.

They usually present a painless clinical picture and evolution towards multiple myeloma is rare. The opposite occurs with secondary extramedullary plasmacytoma in multiple myelomas, which are indicative of aggressiveness and worse prognosis. Malignant neoplasms originating from the B-series immunocytes (B-lymphocytes and plasma cells) include: a) multiple myeloma and solitary plasmacytoma, b) Waldenström macroglobulinemia and other monoclonal gammopathies, and c) chronic lymphoid leukemia and malignant lymphomas. The solitary plasmocytomas (PS) are localized proliferations of plasma cells that can appear in any place where there are reticulo-endothelial system formations. These are infrequent tumors, which comprise between 5-10% of plasma cell neoplasms. They are classified as solitary bone plasmacytoma (POS) and extramedullary plasmacytoma (PEM).

Plasma cell tumors are histologically indistinguishable (MM, POS, PEM) and differentiated based on a combination of specific histological tests and study of systemic involvement. It is important to make a good diagnosis, since the treatment and survival are different.MM is a fatal disease with an average survival of 2-3 years. In plasmacytomas, the prognostic indicator is progression to MM, which occurs in 50% of patients diagnosed with solitary plasmacytoma between 3 and 5 years after diagnosis. For this reason, these patients, despite having had a good response to treatment with radiotherapy and / or surgery, must be closely monitored due to their high progression to MM. For some authors, MM, PSO and PEM represent different manifestations of the same continuous disease and distinguishing one from the other has significant implications for treatment and evolution or survival. The extraosseous involvement of myeloma is infrequent; It has been found in 5% of cases and has been associated with a more aggressive course. This extraosseous involvement, reported in the literature, occurs in various soft tissues. The most commonly reported occurs in the submucosa of the upper airways, although it also occurs in the mucosa of the digestive tract, as well as in the lymph nodes associated with it. Other sites that have been found include the testicle, liver, small intestine, pancreas and colon, there are even reports of spread to the skin.

In the series reported it has been shown that the appearance of extraosseous myeloma usually occurs in several organs and with a more aggressive course; however, no specific genetic modifications have been found that explain this behavior. No specific radiological patterns have been identified that allow an accurate etiological diagnosis to be made from the images.

Case Report

This is a 75-year-old male patient who started three months ago with edema of the right pelvic limb after prolonged standing, to which periods of chronic constipation, asthenia, lost adynamia of three kilograms in a month were added. As an antecedent, it has a pacemaker: Biotronik, Axios SR, for a tachyarrhythmia history. In March 2016, he went to the emergency department where Doppler ultrasound protocol was performed, discarding lower limb vascular compromise. He requested a single-phase abdominopelvic tomography which revealed a homogenous lobulated mass with soft tissue attenuation of 15 x 10 cm retroperitoneal, below of the aortic bifurcation occupying the pre sacral space and involving iliac vessels on the right side (Fig. 1). In the PET / CT study with 18F-FDG, the presence of a retroperitoneal lymph node conglomerate of 17x11cm was reported, which enveloped the vascular trajectories, obliterated and displaced the right ureter causing right uretero-pyeal ectasia, associated with diffuse metabolism in relation to already known primary neoplastic activity. (Fig. 2).At the physical examination, soft, depressed abdomen, no megalia can be felt. Mild edema of the right pelvic extremity. There was no pallor or jaundice. Laboratory tests revealed hemoglobin (HB) of 11.4 gms / dl, total white blood cell count and platelets were normal. The ratio albumin / globulin 1.3, globulin 3.3, tests of renal function, liver enzymes, serum electrolytes, calcium and phosphorus within normal limits.Serum electrophoresis reported: serum levels of immunoglobulins within normal ranges. The urinary proteins of Bence Jones were negative. Bone marrow aspiration and biopsy were normal. He underwent a laparotomy biopsy on May 6th and was reported as a plasma cell neoplasm, which restricts for lambda light chains (Fig. 3). He received treatment with 50 Gy radiotherapy in 25 fractions to the lumbar and pelvic region, and then received 6 cycles of carfilzomib-lenalidomide-dexamethasone (30 mg, 25 mg orally, 40 mg iv, respectively) until February 2017, subsequently received maintenance therapy with lenalidomide 15 mg orally every 24 hours for 21 days as it achieves complete remission of the disease endorsed by PET-CT of February 2017. In October of the same year it refers to a condition of at least 2 months of evolution characterized by horizontal diplopia. I have an

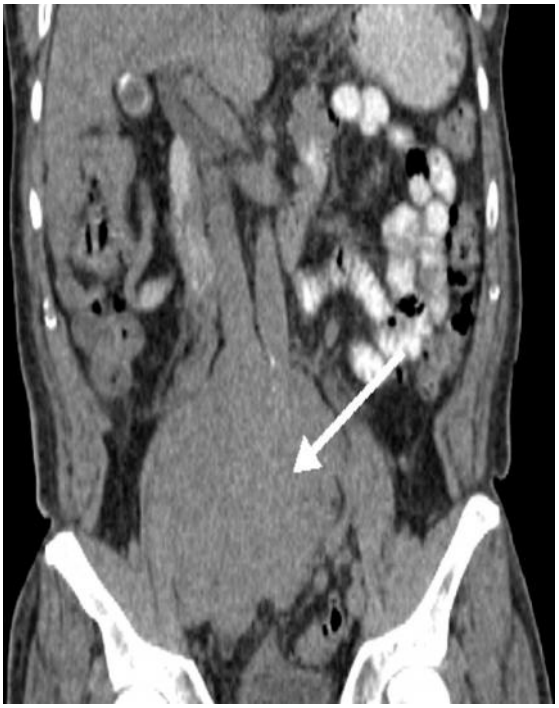

Fig. 1. Abdominopelvic computed tomography: a mass of 8.8 cm located in retroperitoneum.

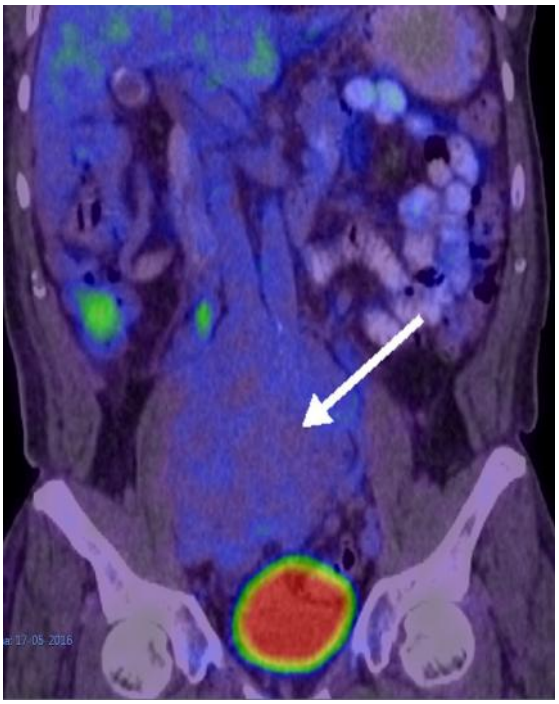

Fig.2 PET-CT retroperitoneal mass with mild diffuse hypermetabolism, with SUV max. 2.0.

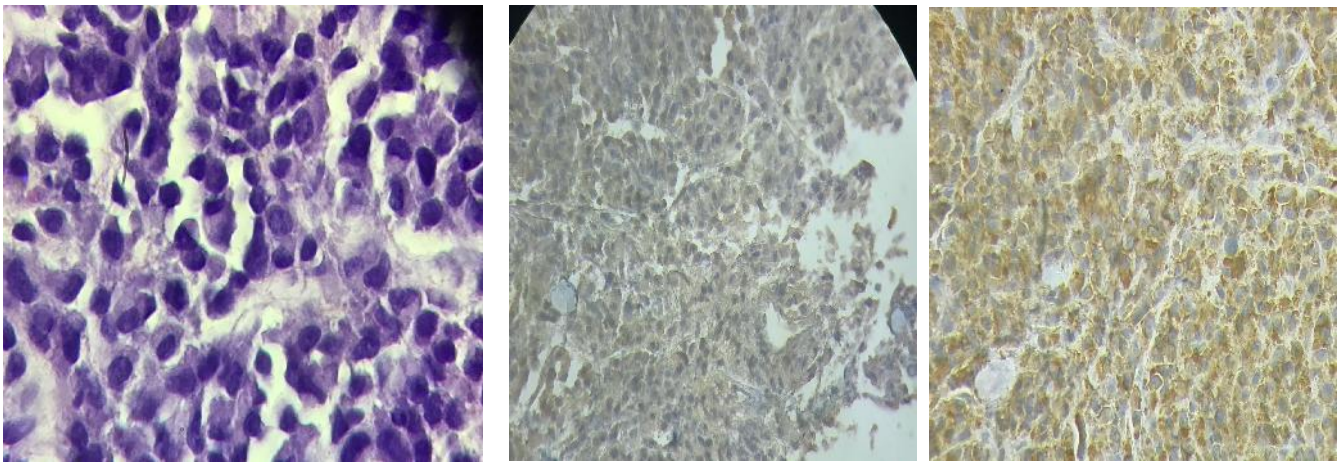

Fig. 3 a) abnormal plasma cell with Large oval and peripheral nucleus b) Plasma cells negative Kappa chains c) Lambda positive chains plasma cells

exacerbation of the symptoms in recent dates, which conditions gait disorders consisting of lateropulsion, ataxic gait. Negates headache, nausea, vomiting, drowsiness or some other symptomatology. Cranial nerves with slow pupillary response, engine motor reflex abolished, consensual present. It does not perform ocular supraversion movements. Force 5/5, ROTs ++ / ++++. Absence of abnormal movements. Other pathological signs are not evoked.The PET / CT study with 18F-FDG in abdomen with retroperitoneal ganglionic conglomerate involving renal vascular pathways and right ureter causing mild ectasia of the ipsilateral pyelocalicial system, mesenteric and pelvic adenopathies with diffuse mild metabolism in relation to recurrence of already known primary neoplastic activity. (Fig. 4).Skull with nodular image in third ventricle without increase in glycolytic metabolism not present in previous studies, to rule out secondary deposits of known primary in the first instance. (Fig. 5). Neurological assessment: patient with Parinaud syndrome or mesencephalic aqueduct secondary to pineal region injury that extends through the pineal recesses to the third third of the third ventricle. Does n ot have magnetic resonance by pacemaker.

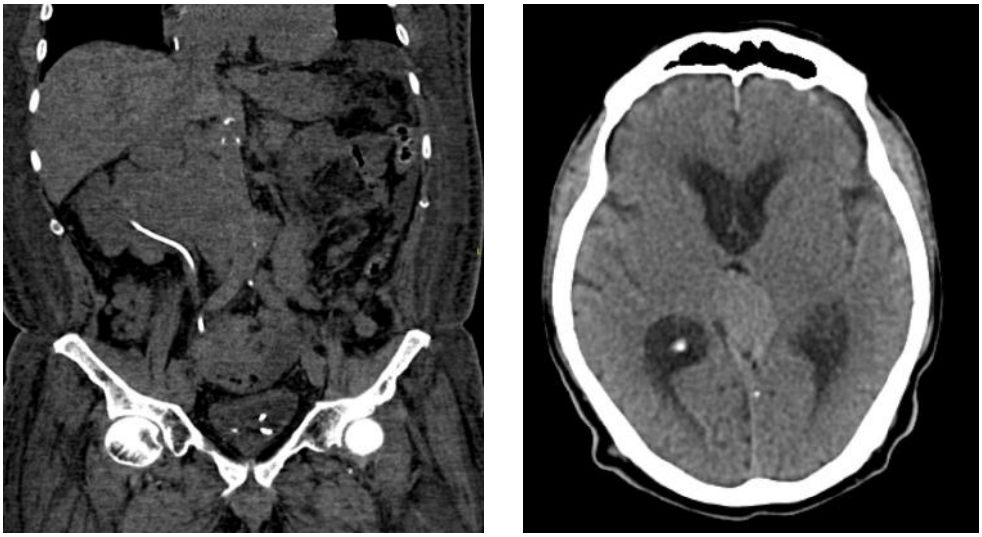

Fig. 4 Retroperitoneal lymph node conglomerate wrapping renal vascular trajectories and right ureterFig. 5 a) Nodular image in third ventricle

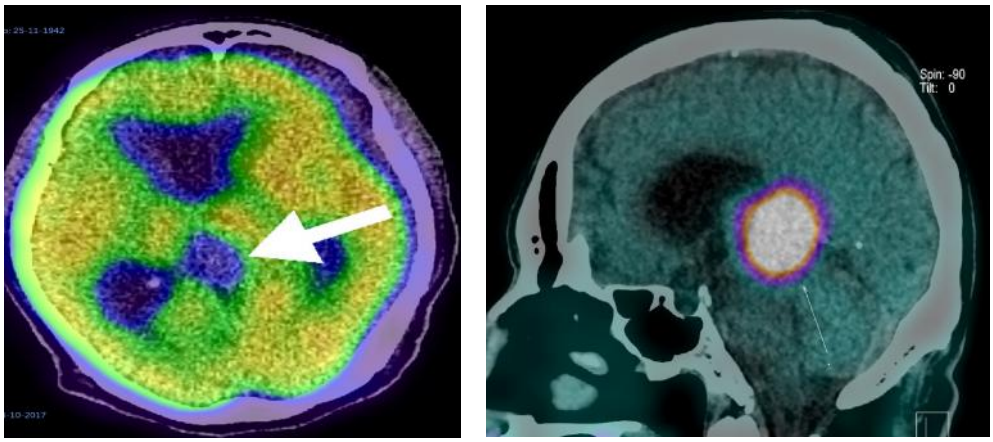

Fig. 5 Nodular image in third ventricle without increased glycolytic metabolism: a) axial plane b) sagittal plane.

Tumor markers (beta fraction of HGC-h, alpha-fetroprotein, carcinoembryonic antigen, CA-19-9, 125 and 15-3 were requested in order to rule out the possibility of a germ cell tumor being normal.) Biopsy of the third ventricle was performed. finding "neoplasm of plasma cells in the central nervous system light chains and light chains negative Kappa and lambda intense and diffusely positive in the cytoplasm of the lesional population: CD138 (+), MUM1, MUN 1 (+), CICLINA D1 (+). second-line chemotherapy with daratumumab-lenalidomide-dexamethasone three cycles, as well as 30 Gy radiotherapy in 18 fractions directed to the CNS In February of 2018, tomography was performed, finding absence of lesion in the third ventricle seen previously, but beginning with renal failure obstructive that required placement of bilateral JJ catheter and with light chain nephropathy clinical evolution of the patient was not satisfactory. He was not considered a candidate for radiotherapy in retroperitoneal injury by previous radiotherapy and risk of injury to the kidney. The third cycle was complicated by severe neutropenia, septic shock due to right pyelonephritis secondary to Klebsiella blee kpc. The laboratories were: HB 10.5, Leu: 4400, Plt: 80 thousand, Nt: 2600, Creatinine 2.5, BUN 30, Normal electrolytes, B2 microglobulin> 4, low immunoglobulins. It also started with radiculopathy because of all the above, they determined progression of the abdominal plasmacytoma and candidate for palliative chemotherapy with dexamethasone and pomalidomide.

Discussion

In the affection of primary lesions to retroperitoneum we must remember that 70 to 80% are neoplasms of malignant nature and represent 0.1% to 0.2% of all the malignancies of our body. Non-invasive imaging techniques help us to characterize them (shape, size, calcifications, septa, thickening of the wall, fat content).The tomography helps us to better define the calcifications, the magnetic resonance for a better contrast between the soft tissues and the ultrasound characterize the vascular invasion. Retroperitoneal sarcomas constitute 0.1% -0.2% of all malignant neoplasms are common in people of the fifth and sixth decade of life, neurogenic tumors 10% -20% occur in young people and have a better prognosis, germ cell tumors extragonadal 1% -2.5% more common in men, Among the hematological neoplasms, lymphoma is the most common retroperitoneal neoplasia occurs in 33% of cases and extramedullary plasmacytoma in 3% -4% of cases. Plasma cell neoplasms can occur as multiple myeloma or as solitary plasmacytoma.

Confirmation of solitary plasmacytoma is only possible when the results of complementary tests: Bence Jones protein, bone x-rays, peripheral blood smear, CSF and plasma Ig and bone marrow biopsy are negative, demonstrating non-recurrence and non-dissemination to the make periodic checks. Multiple myeloma is a malignant clonal neoplasm derived from plasma cells, which results in the overproduction of monoclonal immunoglobulins (ie, electrophoretically and immunologically homogeneous). These plasma cells initially grow in the bone marrow and, subsequently, may invade the adjacent bone or spread

out at a distance; however, its typical presentation is bone lesions secondary to the proliferation of tumor cells and the activation of osteoclasts that cause bone destruction, 99% of patients have M protein in the serum, in the urine or both. In 55% of cases, the M component is IgG; in 25%, IgA, and rarely IgM, D or E. In 20% there is only Bence Jones proteinuria without serum M component. The extraosseous involvement of myeloma is infrequent; it has been found in 5% of cases and has been associated with a more aggressive course; no specific genetic modifications have been found that explain this behavior. PEM is a rare entity of plasma cell tumors, which accounts for less than 5% of all of them. However, 15% of PEMs can evolve to a generalized form, such as MM, which has an annual incidence of 5 cases per 100,000 inhabitants. MM usually affects only the bone marrow, although 20% of cases may present a lesion in other locations. PEM is defined by the existence of a clonal focus of plasma cells outside the bone marrow, generally of the IgA or IgG type and light chains K, without evidence of organic involvement (absence of bone lesions, anemia, hypercalcemia or renal failure), of component M (although up to 25% of cases may be present in low values) or of spinal cord involvement (plasma cell count after bone marrow biopsy <10%). It usually appears more frequently in males, most of the cases between the fourth and seventh decades of life. Its most common location is the upper aerodigestive area (nasal cavity, paranasal sinuses, hypopharynx) and represents about 90% of cases. The remaining 10% present a great variability of locations, mainly cutaneous, urological and, above all, digestive. variable: from completely asymptomatic forms to others that present as digestive hemorrhages or abdominal pain, and even as an intestinal obstruction.

Once diagnosed, it is important to rule out that it is a manifestation of MM, which can occur in 4-5% of cases. This is important, both from the point of view of treatment since in the PEM the treatment in most cases is local, while in the case of MM it is necessary to add systemic treatment with chemotherapy as well as the prognosis, much worse in cases of MM with extramedullary disease (less than 24 months after diagnosis). The treatment of PEM depends on its location, size and histological grade. The prognosis of these tumors is usually good, and survival exceeding 65% at 10 years is achieved. Recurrence, if it occurs, is usually located in the lymph nodes or locally; Distant metastases are rare. They usually appear after 2-3 years of follow-up.

In addition to its possible appearance, lifelong follow-up of these patients is important, since about 15% will develop MM (in solitary plasmacytoma bone this occurs in up to 50% of cases), even up to 15 years after your diagnosis. The clinical presentation presented our attention due to its evolution after the intervention, since tumor recurrence appeared very early, producing a rapid deterioration without response to chemotherapy in abdominal plasmacytoma, however good response to treatment in cerebral plasmacytoma .

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