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Frontal Right Plasma Cell Granuloma; Case Report

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ABSTRACT

The Plasma Cells Granuloma is a relatively rare lesion, that forms a nodule or a mass, it is conformed of polyclonal plasmatic cells in storiform background fibrosis and spread of fusiform cells. We present the case of a 62-year-old woman, with a history of oophorectomy and increased volume in the right frontal area of the head, then presenting neurologic signs of frontal lobe syndrome. Radiologic images showed a wide right frontal lobe neoplasia that spreads diffusely. Histologically the tumor was formed by plasma cells, B lymphocytes infiltrate, and numerous blood vessels. It was positive for CD20, kappa, lambda, CD3, and CD4. Even though it is uncommon, it can develop at any place and must be included in the differential diagnoses list for plasma cells neoplasia. The positivity of light chains kappa and lambda make evident the polyclonality that confirms the diagnosis.

Keywords

Inflammatory myofibroblastic tumor, Inflammatory pseudo tumor, Brain tumor, Plasma cells granuloma, Plasma cells tumors.

Introduction

Plasma Cells Granuloma (PCG) is a benign tumor that affects any part of the body, it has been reported at many sites in multiple or single forms [1]. The plasma cell granuloma has received a diversity of names [2], among which stand inflammatory pseudotumor [3].

Several other synonyms have been used for PCG that includes myofibrohistiocytic inflammatory tumor, inflammatory pseudotumor, histiocytoma, fibroxanthoma, xanthomatous pseudotumor, myxoid hamartoma, lymphoid hamartoma, fibrous xanthoma, benign myofibroblastoma, pseudosarcoma, and more recently, inflammatory myofibroblastic tumor [2].

PCG is an uncommon disease that can affect any stage of life, but it is more frequently in childhood and young adults, with a slight prevalence in the female gender [1]. Clinical and radiological manifestations are not characteristic, it presents as a nodule or tumors that may appear in any organ, and the symptoms depend on the localization and size of the lesion [2].

PCG is a benign tumor or pseudotumor and is also called fibro inflammatory or inflammatory pseudotumor, histiocytoma, and fibro xanthoma [2]. Here we present a rare case of intracranial plasmatic cells granuloma in the right frontal lobe of the brain, that was diagnosed initially as a sarcoma according to the extended radiological images.

Case Report

A 62-year-old woman, without relevant medical history, had only oophorectomy 20 years ago. The patient initiates her condition in May 2017 with a sensation of increased volume of the right frontal area of the head, so she was valued by a physician, who diagnosed a lipoma, and received symptomatic treatment. One month later, mental abnormalities and difficulty to articulate words were referred by the daughter´s patient. In August 2017 holocraneal intense pulsatile headache was added, accompanied by weakness of lower limbs, instability for walking causing several falls, emotional lability, easy crying, and low tolerance to

frustration. Later she entered the NINN. A head CT scan showed osseous destruction, hyperdense and ill-defined image in the right hemisphere with displacement of the middle line. MRI images showed a meningeal injury that affects soft tissues and the frontal lobe (Figure 1a-1c). The initial diagnostic was probable sarcoma and for that reason the patient underwent surgery. Blood tests routine, viral antibody, electrolytes, liver and renal function tests, coagulation tests and other tests results were in normal parameters.

Three different tissue specimens were sent to pathology: 1- Irregular meningeal sample, of 30x30x30 mm, grayish-white in color, showing a finely granular surface, friable, infiltrating the meninges diffusely. 2- A fragment of dura mater, of 12x10x10mm (Figure 1d), white, rubberized appearance, firm, with a diffuse elevated granular lesion of 60x50x20mm in the central zone (Figure 1e). This lesion was yellowish grayish, nodular, granular, soft, despicable, friable that infiltrates the rest of the meninges in a nodular form. 3- An osseous fragment of the frontal bone, of 12x10cm, with partial detachment of it (Figure 1f). On one side it is appreciated a granular-sandy lesion, yellowish-brown, ill-defined, that infiltrates diffusely the thickness of the bone, it measured 50x30mm. Histologically a dense lymphoplasmacytic infiltrate is appreciated (Figure 2a), with predominant normal plasma cells with hyaline areas (Figure 2b), and discretely thickened non-hyalinized vessels (Figure 2c). Immunohistochemistry staining was performed, with high positivity for CD20 (Figure 2d) and CD78 (Figure 2e). The light chains kappa and lambda were positive, and focally immunoreactive for CD3 (Figure 2f), CD4, and CD68, the Ki67 was negative. Epstein Barr virus (LMP-1), herpesvirus 8, and IgG4 were also negative. It was diagnosed with Plasma Cells Granuloma.

Discussion

Plasma Cells Granuloma is characterized by the formation of granulomatous masses of different sizes, located in various organs. Clinically, PGC occurs as any tumor is benign or malignant in any anatomical site. It can be associated with trauma, previous surgeries, prolonged inflammatory processes, infections, such as herpesvirus, Epstein-Barr virus, and even IgG4 which in our case were negative [4].

Over the years, since it was first described, mechanisms of action and new diseases have been added in association. It should be considered as a malignant tumor because poor prognosis cases have been reported, with frequent recurrences, malignant transformation, and even high morbidity. Mandelbaum, et al. [5], have drawn attention to the possible association between plasmatic cells granuloma and carcinoma. This requires a cautious assessment of

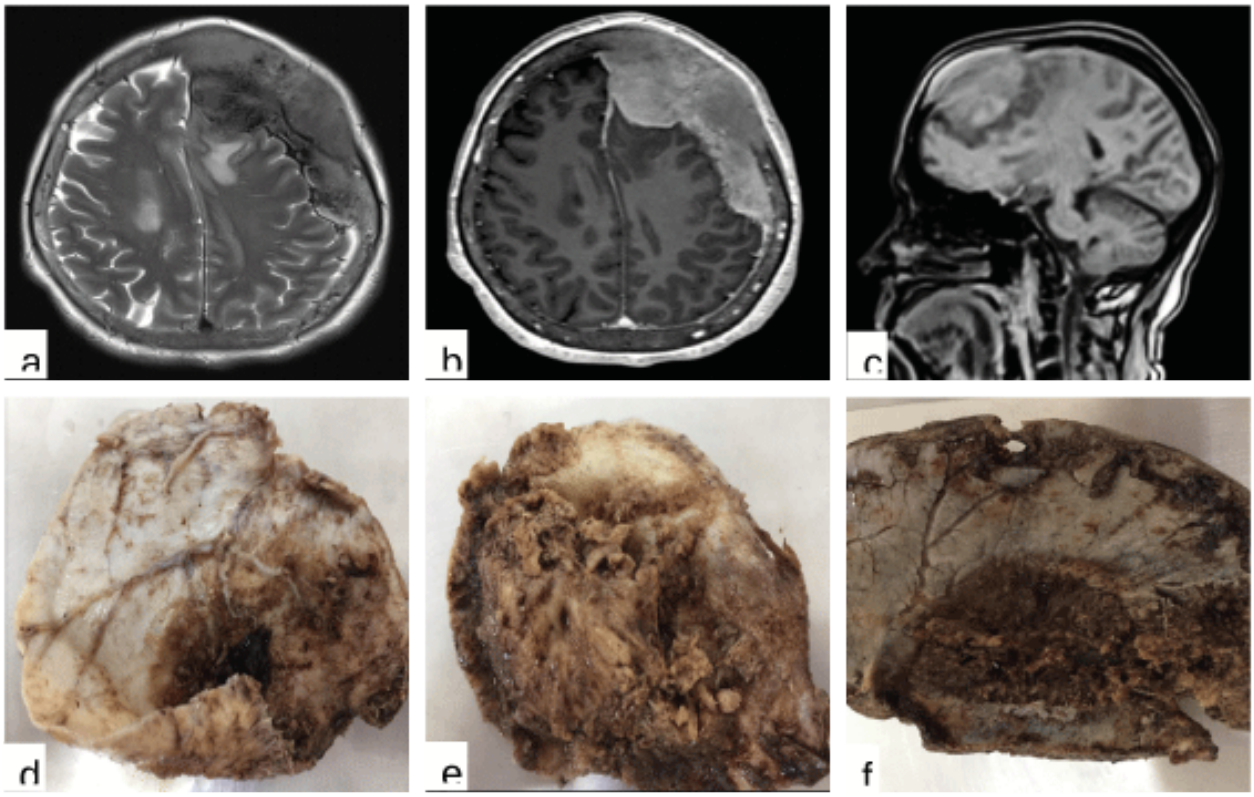

Figure 1: Shows the radiological images of the lesion. (a) MRI in T1 coronal section shows the intra and extra dural meningeal lesion. (b) MRI in T2 coronal section shows a hyperintense heterogeneous lesion. In (c) Sagittal section we observed nodular lesion with extensive perilesional edema. (d) Macroscopic appearance of the lesion, which corresponds to an irregular segment of dura mater on the external part that measured 12x10x10mm, diffusely infiltrated by a clear to yellowish-brown neoplasm and in (e) through the convexity this was white with a rubbery appearance, firm, and in the central portion there was a diffuse lesion raised granular clear coffee that measured 60x50x20mm, and in (f) we observed fragment of calotte of the frontal region that shows an infiltrating neoplasm, high, gray-brown, granular and soft.

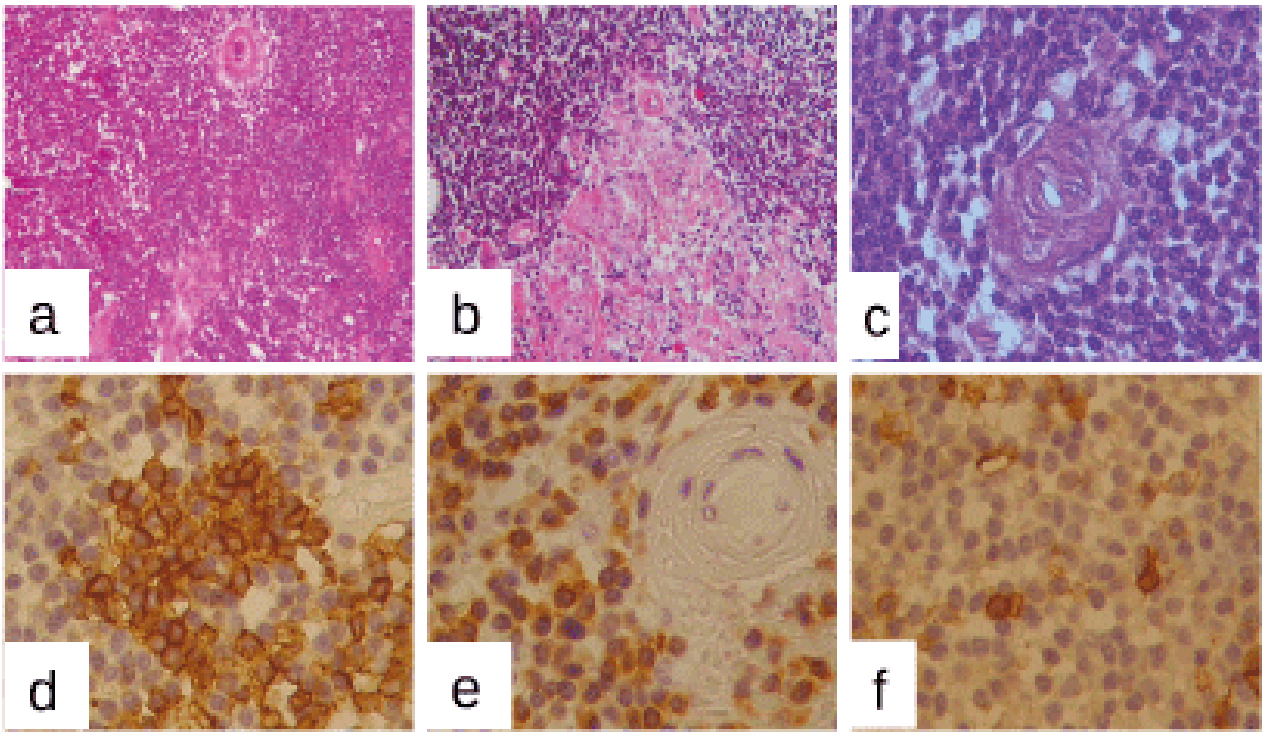

Figure 2: Histologically the lesion was formed by a dense infiltrate of plasma cells and lymphocytes in (a) H & Ex100), focally hyaline and collagenized proliferation is seen in (b) and a closer approach we observed better homogenous plasma cells and focally hyalinized vessels (H & Ex40). With immunohistochemical staining we observed positivity for CD20 in (d). (e) CD78 was positive plasma cells in (f), and CD3 focally immunoreaction positive immunoreaction (IHx400).

the clinical, radiological, histological, and immunohistochemical manifestations. The diagnosis is made by exclusion and mainly by pathological examination of the lesion [1].

Recently the WHO classification of soft tissue tumors classified them into three types: Myofibroblasts pattern freely organized in a myxoid edematous background, showing plasma cells, lymphocytes, eosinophils, and blood vessels, which shows dense aggregates of fusiform cells arranged in a myxoid stroma, or collagenized, mixing with a distinctive inflammatory infiltrate and diffuse groups of plasma cells and lymph nodes. Tumor with a predominance of collagen fibers, which resemble scar tissue, with the presence of plasma cells and scattered eosinophils, with Castleman disease as a differential diagnosis [5,6]. This variant may have cytological atypia with nuclear pleomorphism and increased mitotic activity; these characteristics are rare and may be associated with malignant transformation [5]. Our case, due to its histological characteristics and the presence of abundant blood vessels, could be included within the category of Myofibroblast pattern freely organized in an edematous myxoid background, showing plasma cells, lymphocytes, eosinophils, and blood vessels [6].

PCG is not a difficult diagnosis to make, normal plasma cells are found, although they become atypical, with discrete cellular pleomorphism, there may be a dense lymphocytic infiltrate, predominantly B lymphocytes (CD20+), T lymphocytes are also observed in positive immunoreaction for CD3+, CD4+, and CD8+, as well as macrophages (CD68+), dendritic cells (Fascin +), eosinophils and above all it is striking that both kappa and lambda light chains are positive because it is a polyclonal disease [1,6].

As this entity can be confused with a plasma cell neoplasm, surgical pathologists should consider complementary studies to evaluate the clonality of plasma cell proliferation, especially during the intraoperative consultation. Although the etiology of these lesions is not clear, recent literature and immunohistochemically stains suggest that PCG is within the spectrum of IgG4-related diseases, which would have a clinical significance that would affect the treatment and the associated potential disease in the distant body sites [4].

The differential diagnosis will be made with chronic infections, such as syphilis, fungal infections, benign neoplasms, reactive granulomas, foreign body granulomas, and malignant neoplasms depending on the location of the lesion, as well as solitary plasmacytoma, plasma cell dyscrasias, and if there is multiple conditions with myeloma [6-8].

Inflammatory pseudotumor (IPT) is a heterogeneous group of lesions that occur in various organs, histologically characterized by fibroblastic and myofibroblastic proliferation with inflammatory infiltrate [1,2]. The inflammatory myofibroblastic tumor (IMT) is a neoplastic counterpart of ITP [7], which shows an aberrant expression of ALK and its gene translocation; however, the presence of IgG4 positive plasma cells may be useful for the

differential diagnosis between MTCT and sclerosing disease related to IgG4 [4].

Approximately 50% of conventional inflammatory myofibroblastic tumors (MTCT) harbor the rearrangement of the ALK gene and over express ALK. Recently, gene fusions involving other kinases have been implicated in the pathogenesis of TMI, including ALK, ROS1, PDGFRB, NTRK1 and RET [9]. Castleman disease (CD) is a rare lymphoproliferative disorder that is characterized as either unicentric that has an excellent prognosis and the multicentric type that is a systemic disease that most commonly occurs in the context of HIV infection and is associated with human herpesvirus 8 [10]. It is a non-clonal lymphoproliferative disorder and one of the most common causes of non-neoplastic lymphadenopathy. The spectrum of Cattleman's disease includes the classic and well-recognized hyaline vascular type, the type of plasma cells, and the multicentric types of a wider histological range [6]. We must consider it with or without affection to lymph nodes and the predominance of plasma cells and vascular damage and/or vascular proliferation and proliferation of lymphocytes that reach germinal centers that resemble the lymph node [6].

The biological behavior of PCG is variable, and the treatment consists of radical surgical removal or destruction by electrosurgery and curettage, or electrosurgery associated with cryosurgery [4,5]. Radiosurgery can also be used, as well as chemotherapy, corticosteroids, and immunosuppressants [5]. The prognosis remains uncertain, there are cases in which it acts as a reactive inflammatory process, in others as a benign tumor, but there are also reports of cases with malignant neoplastic behavior at least locally with obvious infiltration and destruction of the affected tissues [10].

Conclusions

Plasmatic cells granuloma is a relatively rare entity, with benign histological characteristics, which can have a very aggressive local behavior. A definitive diagnosis is reached by pathological study of the lesion. It should be present in the differential diagnosis before any expansive and osteolytic lesion.

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