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A rare case of solitary plasmacytoma presenting as a skull lesion

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Abstract
Plasmacytoma is a neoplastic proliferation of monoclonal plasma cells in bone or soft tissue, potentially indicating multiple myeloma. Solitary plasmacytoma is rare, with an incidence of 0.15/100,000, commonly affecting vertebrae and skull bones or the head and neck region. Etiology includes chronic stimulation, radiation, viral infections, and genetic factors. This case involves an 82-year-old male with a painless parietal swelling, revealing an osteolytic lesion upon imaging. Histopathology showed pleomorphic plasma cells, confirmed by CD138 positivity on immunohistochemistry, leading to a plasmacytoma diagnosis. Comprehensive diagnostics are crucial for accurate identification and treatment.

Keywords: Plasmacytoma, monoclonal, histopathology, osteolytic, CD138, diagnosis.

Introduction
Plasmacytoma is a tumour of neoplastic monoclonal plasma cells proliferation in either bone or soft tissue [1]. Solitary plasmacytoma may be an isolated tumour in any part of body and may be the first manifestation of a Multiple myeloma. [2] Solitary plasmacytoma is a rare condition with an incidence of 0.15/100.000 [3]. Solitary bone plasmacytomas affect most commonly the vertebra and the skull bones. Extramedullary plasmacytomas tend to arise more commonly from the head and neck region, nasal cavity, and nasopharynx [4] however the etiology of solitary plasmacytoma is unknown, chronic stimulation, radiation overdose, viral infections and genetic interaction in the reticuloendothelial system have been suggested to contribute to the development of the lesion [5].

Case report
82-year-old male patient came with complaints of left sided painless parietal swelling. Initially small in size gradually increased in size to 3x3 cms, with the differential diagnosis of Metastasis / Multiple myeloma was given by the clinician. As a routine complete blood count was done with Haemoglobin of 13gms/dl and all other parameters were normal. ESR was found to be raised to 30mm with normal renal function tests.
Ultrasonogram revealed a hypoechoic mass of size 5.2 x 2.2 cms noted in left parietal region with the underlying bone showing increased vascularity and feeding vessel into the lesion. An impression of a well-defined mass of left parietal region with underlying bone erosion was made and was further suggested CECT was made. Further CECT was done and showed an osteolytic lesion in the left high frontal region with soft tissue component and associated multiple lytic lesion diffusely involving the skull was made and a differential of Metastasis / Multiple myeloma was given, suggesting scintigraphy or FNAC for further evaluation. Meanwhile the patient was covered with antibiotics, analgesics and antiepeleptics drugs.
Histopathological examination revealed multiple grey white, irregular, soft tissue mass, largest of size 4.3 x 4 x 1.2 cms, and smallest measuring 3.2x 2.5 x 0.2 cms. Cut surface was whitish, firm in consistency with a tiny bony hard fragment. Microscopically it was a partly encapsulated tumour with tumour cells arranged in sheets and lobules separated by delicate fibrovascular septa with many thin walled blood vessels. Individual tumour cells exhibit moderate to marked pleomorphism with abundant eosinophilic cytoplasm, vesicular nuclei and most showing prominent nucleoli. Tumour cells were admixed with scattered inflammatory infiltrates. Areas of necrosis, fragment mitosis (3/10HPF) along with atypical forms, apoptotic bodies and bizarre giant cells were also seen.

Possibilities of Histiocytic sarcoma, High grade Non-Hodgkin's lymphoma or metatstatic carcinoma was considered.

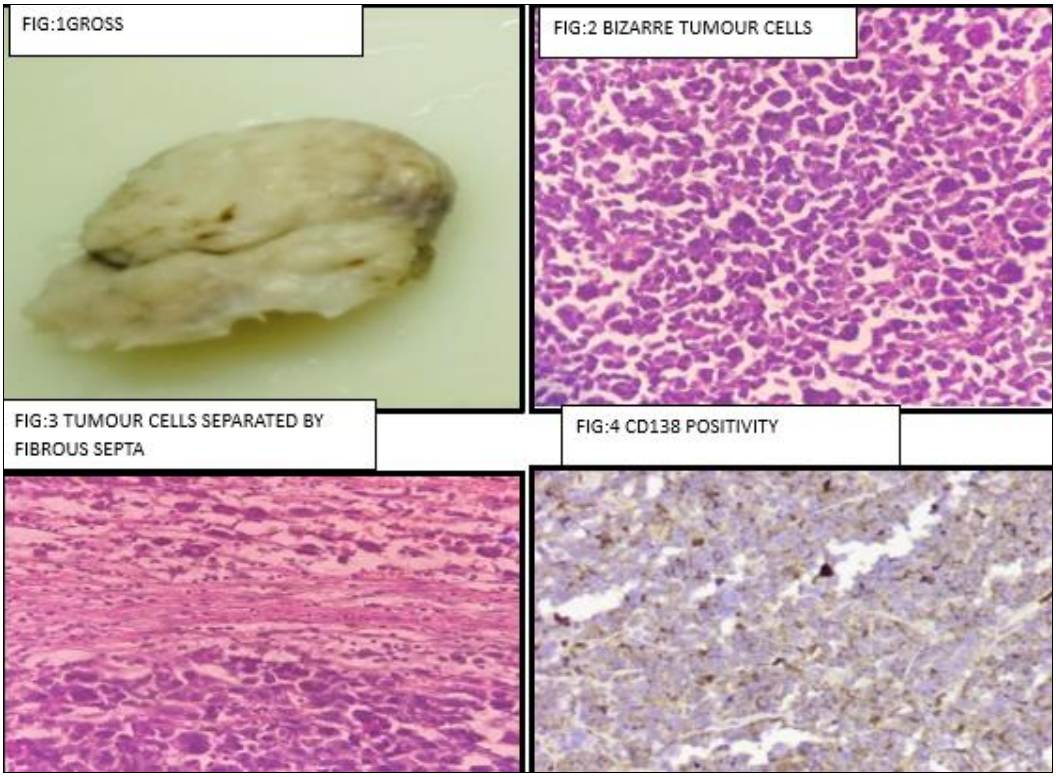
Immunohistochemistry was done using CD38, CD163, Pancytokeratin, S-100 and CD20 was done to confirm the diagnosis. IHC with PanCK, CD20, CD38, CD163 and S-100 were negative except for CD138 which showed moderate cytoplasmic positivity and a diagnosis of plasmacytoma was made.

Immunohistochemically examination was done with antibodies for free light chains and Kappa and lambda was done which showed

Free Kappa - light chain - 5.61gm/dl

Free Lambda - light chain - 2.15gm/dl

Free Kappa/free lambda ratio - 2.60 was given, further confirming the Plasma cell disorder.



Discussion

Solitary plasmacytoma is a rare plasma cell tumour that does not involve the bone marrow. Solitary plasmacytoma of the skull is an uncommon entity that is being characterized by localized proliferation of neoplastic monoclonal plasma cells [6]. The median age of patients at the time of diagnosis of solitary plasmacytoma is 55-60 years, comparatively lower than in patients of multiple myeloma and the male to female ratio varies from 1, 2:1 to 2:1[7]. Recent studies suggest that the interactions between the SPs and their bone marrow microenvironment, as follows, that this might be critical and notably in the early stages of myeloma genesis. The involvement of clonal plasma cells with the bone marrow environment may determine the stage of the SP [8]. The interactions between the clonal plasma cells and their background is also relevant in extramedullary plasmacytomas. The second pathogenic pathway is related to the "occult marrow disease". The clonal bone marrow plasmacytoma representsa minority at diagnosis and therefore, under-diagnosed by imaging and is minimized by bone marrow biopsy immunophenotyping. However, modern diagnostic methodology, includes the evaluation to be routinely performed by multiparameter investigations such as serum / urine free light chain ratio and flowcytometry [9-11]. The third pathogenic process, in relation to the bony lesions might be detected more precisely, using low-dose whole-body CT, the MRI, the FDG-PET and PET-CT. Bone destruction was analysed by an increased osteoclastic resorption and by decreased bone formation. The osteoclastic resorption is considered to be an early event, occurring long before the first symptoms of the disease can be manifested. Tumour plasma cells, as well as the hematopoietic microenvironment produce inflammatory cytokines, osteoclast activator factors, and osteoclast colony stimulating factors [12]. Plasmacytomas can be divided into two groups depending on its location, solitary bone plasmacytoma and solitary extramedullary plasmacytoma. Multiple myeloma (MM) is considered to be the systemic form of the pathology which is characterized by multifocal disseminated lesions, and is primarily based on CRAB criteria such as increased calcium, renal insufficiency, anaemia, or multiple bone lesions, representing the most common tumour of plasma cells[13].SP accounts for about 70% of all cases and tend to occur primarily in red marrow-containing bones[14].Most common osseous location of solitary plasmacytoma are the pelvis, spine, femur, humerus and ribs, in which they may lead to bony destruction [15].Authors recommend certain criterias for the diagnosis of Solitary plasmacytoma or extramedullary plasmacytoma these include

1. Single lytic bone lesion (in case of Solitary plasmacytoma of bone) or extramedullary mass lesion (in case of extramedullary plasmacytoma), that is histologically composed of clonal plasma cells
2. Normal bone marrow aspirate and biopsy without any clonal plasma cell population.
3. Absence of bone involvement, excluding solitary lesion, on skeletal survey or MRI study of spine and pelvis

4. Absence of end organ damage, such as CRAB criteria that is attributable to plasma cell dyscrasia [16].

Histopathological examination of plasmacytoma is straightforward, and except for cases where the tumour cells are poorly differentiated. Microscopically the neoplasm consists of plasma cells of variable degree of plasmacytic maturation. Mature cells usually have clumped nuclear chromatin, abundant cytoplasm, and no nucleoli. Immature cells often have large nuclei with prominent nucleoli. Some cases may also have multinucleated and pleomorphic plasma cells.

Due to the rarity of the disease, there have been no randomized studies about the best treatment options, and data reported in the literature are considered to be controversial [7]. Yet the recommended treatment with radical radiotherapy, with a margin of at least 2 cm and a dose of 40 Gy in 20 fractions and for SBP >5 cm, a higher dose of up to 50 Gy in 25 fractions should be considered [17].

Conclusion

In conclusion though a rare entity, a bony lytic lesion of any age the possibility of plasmacytoma as a differential has to be considered. Histopathological examination is considered a gold standard, still a panel of investigations is required for its confirmation. The option of treatment for SPs is site specific with either radiotherapy, surgery or a combination of both. The diagnosis of solitary bone and soft tissue plasmacytomas is fairly straightforward and a biopsy is indicated at the earliest in suspected cases of plasmacytoma. The increased risk of transformation of plasmacytomas into disseminated disease necessitates the patient to be under a periodic surveillance [16].

Conflict of Interest

Not available

Financial Support

Not available

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