

Plasmablastic Myeloma in a Young Patient Presenting With an Intracranial Mass Lesion - A Rare Case Report

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ABSTRACT

Plasma cell dyscrasias are a heterogeneous group of diseases which are characterized by the expansion of monoclonal plasma cells that produce monoclonal immunoglobulins. These neoplasms account for 1% of all malignant tumours. They can present as a solitary lesion (plasmacytoma) or with bone marrow involvement (multiple myeloma). The usual age of presentation of these plasma cell neoplasms is in the seventh decade. Occurrence before 30 years of age is very rare. We report a case of a 20-year-old male who presented with an intracranial space occupying lesion which was initially diagnosed as plasmacytoma and with further work up and investigations; he was finally diagnosed with plasmablastic plasma cell myeloma. In the plasma cell dyscrasia category the plasmablastic morphology is very rare. This case is being presented because of the unusual age, location, morphology, nature and sequence of presenting events.

Keywords – Multiple myeloma, Intracranial SOL, Plasmablastic myeloma, Plasmablastic lymphoma, *Meningioma, Plasmacytoma*

INTRODUCTION

Plasma cell neoplasms account for 1% of all malignant tumours. They include multiple myeloma, solitary plasmacytomas and syndromes which are caused due to tissue immunoglobulin deposition.^[1]

They are characterized by the neoplastic proliferation of a single clone of plasma cells which can present as a single lesion (solitary plasmacytoma) or as

multiple lesions (multiple myeloma, MM).^[2] Solitary plasmacytomas can arise within soft tissues where they are referred to as solitary extramedullary plasmacytomas or they may involve the bony skeleton where they are called solitary bone plasmacytoma. The common sites of involvement of solitary extramedullary plasmacytomas include the head and neck regions, upper aerodigestive tract, gastrointestinal tract, bladder, central nervous system, thyroid, breast, testes, parotid gland, lymph nodes and skin. Extramedullary plasmacytomas can arise in patients with multiple myeloma at any time during the course of the disease and are associated with a poor clinical outcome.^[3]

Plasmacytomas involving the CNS are very rare.^[4] These extramedullary plasmacytomas are referred to as intracranial plasmacytomas.^[5]

Intracranial plasmacytomas in young individuals have been very rarely reported. Here we present such a rare case of a 20-year-old male who presented with an intracranial space occupying lesion which was initially diagnosed as plasmacytoma and with further work up and investigations; he was finally diagnosed with plasmablastic plasma cell myeloma.

CASE REPORT

A 20 year old male patient presented to the neurosurgery OPD with a history of on and off fever, headaches and seizures since 3 months. He also complained of right lower limb weakness and a gradually

developing scalp swelling since last 2 months. No changes in personality were noted by family and friends. On examination the patient was conscious and well oriented to time, place and person. The pupils were normal in size and were reactive to light (GCS – E4V5M6). Power in right lower limb was 4/5. The deep tendon reflexes were brisk and extensor response was seen on eliciting the planter reflex. A swelling was observed on the left side of the scalp measuring 5X3 cm in size. The swelling was firm in consistency with restricted mobility and was non-compressible. It was non-tender with unremarkable overlying skin. Rest of the systemic examination was within normal limits.

Non-contrast computerised tomography head showed the mass involving the cranial vault firmly attached to the dura. There was slight compression of the adjacent brain parenchyma. Rest of the preoperative investigations were unremarkable. Based on the clinico-radiological suspicion of meningioma, the patient underwent left frontal craniotomy with wide local excision of the mass the adherent dural layer along with cranioplasty. Histopathological examination showed presence of a tumour composed of plasma cells which were positive for CD45, CD138, CD56, mum1 with lamda restriction. Kappa was negative. A histopathological diagnosis of plasmacytoma was given. The patient was referred to haematology department for further work up and management.

On routine hematology, he was anaemic with peripheral smear showing mild anisocytosis with marked rouleaux formation (figure 1). There was mild leucocytosis with myeloid left shift. Platelet count was normal.

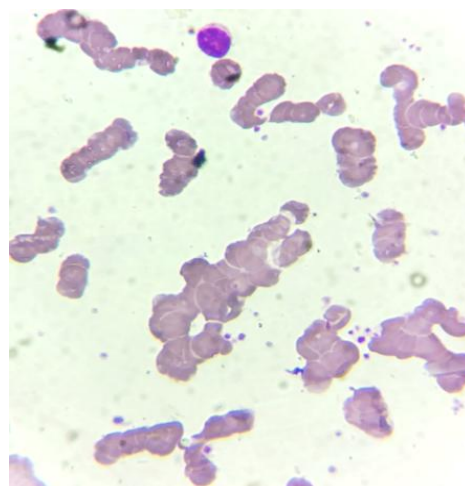


Figure 1: Rouleaux formation on PS

Serum IgA levels were elevated (39g/Lt, reference 0.57-5.34). IgG and IgM levels were reduced (IgG = 5.98g/Lt reference 6.5-16.2g/Lt, IgM = <0.17g/Lt reference 0.30-2.65g/Lt). Serum lambda free light chain was high (303mg/L, reference 5.71- 26.3). kappa/lambda ratio was reduced (0.030, reference 0.26-1.65).

The bone marrow aspirate showed hypercellular marrow composed of clusters and sheets of atypical plasmacytoid cells (Figure 2). Many of the plasma cells were immature and showed anaplasia with large hyperchromatic irregular nucleus and prominent nucleoli. Frequent mitosis was identified.

In biopsy these anaplastic cells were seen replacing the normal hematopoietic cells of the bone marrow (Figure 3). They showed irregular enlarged nucleus with vesicular chromatin and prominent nucleolus. These cells had scant amount of cytoplasm and no perinuclear hof. On immunohistochemistry these cells were positive for CD45, CD117 and showed lambda restriction (Figure 4). Kappa was negative. CD 34 was negative in tumour cells but showed presence of increased angiogenesis in the tumour cell clusters (Figure 5). Based on clinical, serological and bone marrow findings, a final diagnosis of plasmablastic plasma cell myeloma was given.

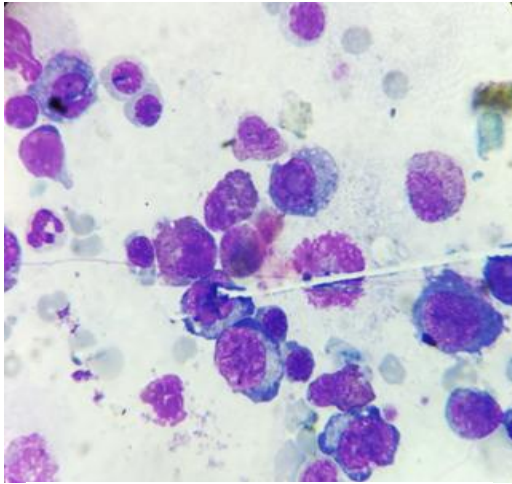


Figure 2: BMA showing atypical plasmacytoid cells

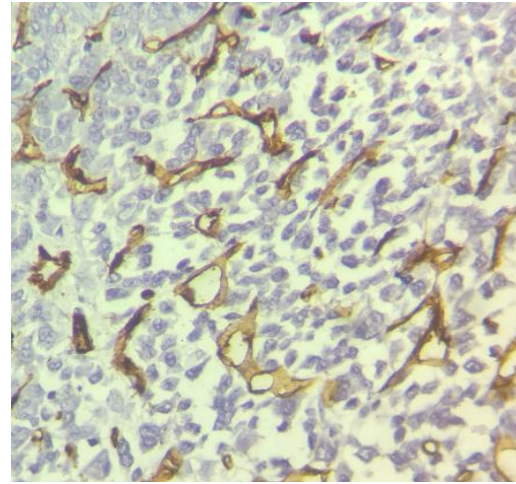


Figure 5: CD 34 showing presence of increased angiogenesis

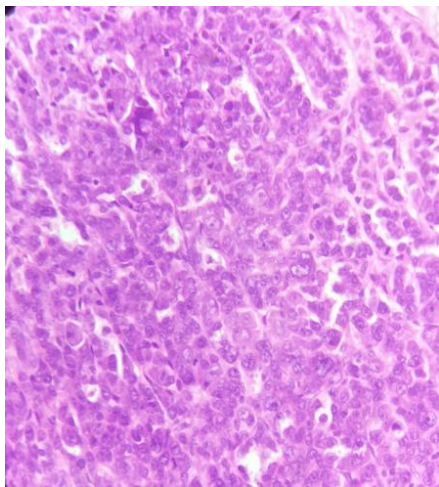


Figure 3 : Bone marrow biopsy showing anaplastic plasma cells replacing the normal bone marrow hematopoietic cells

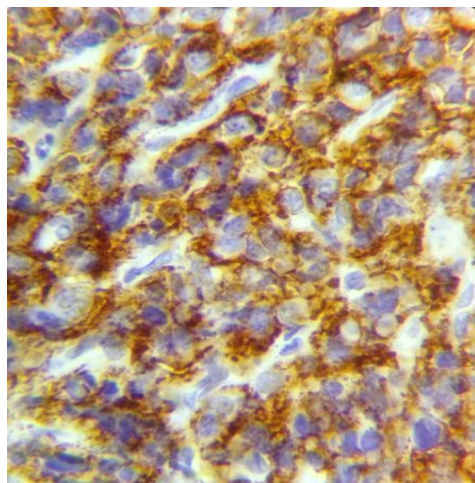


Figure 4 : IHC showing lambda chain restriction

DISCUSSION

Multiple myeloma (MM) is a neoplastic plasma-cell disorder that is characterized by clonal proliferation of malignant plasma cells in the bone marrow, monoclonal M protein in the blood or urine, and associated organ dysfunction. It accounts for 1% of all malignancies and is the second most common haematological malignancy after lymphoma.^[6] The median age at diagnosis is approximately 70 years.^[7] Less than 2% of patients are younger than 40 years of age at diagnosis and it is still rarer in patients younger than 30 years.^[8]

The cytomorphic features of plasma cell tumours ranges from mature, atypical to plasmablastic or anaplastic plasma cells. Plasmablastic multiple myeloma is a morphologic subset of myeloma, in which the bone marrow aspirate/ bone marrow biopsy shows $\geq 2\%$ of plasmablasts. Plasmablasts are the most immature form of plasma cells. Plasmablastic myeloma represents 5-15% of the cases of multiple myeloma.^[8] Plasmablastic morphology of the neoplastic plasma cells is an independent predictor of poor survival, median survival of these patients being 1.9 years.^[10] Therefore, early identification of this aggressive variant of multiple myeloma is necessary for optimal patient management.

A very close differential of plasmablastic myeloma is plasmablastic lymphoma which show significant

morphological and immunophenotypic overlap with plasmablastic myeloma. Plasmablastic lymphoma is composed of cells resembling immunoblasts admixed with other cells with more mature plasmacytic differentiation. Although it is a B cell lymphoma, it expresses plasma cell markers CD138, CD38 and MUM-1 and lacks the expression of B-cell markers like CD20 and PAX-5.^[11] Therefore immunohistochemistry is not helpful in differentiating the two entities.

Certain clinical features like osteolytic lesions, diffuse bone marrow involvement and the presence of an M protein favour the diagnosis of myeloma whereas, features like involvement of oral cavity, HIV infection and Epstein-Barr virus association favour plasmablastic lymphoma. Our case had diffuse bone marrow involvement along with the presence of M protein in the serum. He was HIV and EBV negative so a final diagnosis of plasmablastic myeloma was given.

The case was unique in presentation as the patient presented at a very young age (20 years) with an intracranial dura based space occupying lesion (SOL) which on histopathology was diagnosed as intracranial plasmacytoma and on further work up was finally diagnosed as a case of plasmablastic myeloma. This case also highlights the importance of examination of bone marrow biopsy samples in cases which present as solitary plasmacytomas to distinguish them from cases presenting as a dominant mass in a patient with systemic plasma cell myeloma as in our case.

CONCLUSION

Although multiple myeloma is very rare in young age, a high index of suspicion should be kept by the surgeons and pathologists and this entity should be kept as a differential diagnosis in such young cases who present with intracranial lesions as their first presentation. Bone marrow examination is vital to clinch the diagnosis, to differentiate cases of solitary plasmacytomas from those presenting as a

dominant mass in a case of multiple myeloma. Recognition of plasma cell myeloma in young age patients is further necessary as they may have plasmablastic morphology which has an increased risk of complications, relapse and a refractory disease.

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