Extramedullary Plasmacytoma: A Diagnostic Challenge

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ABSTRACT
Myeloma is a cancer of plasma cells. Myeloma. Multiple Myeloma involves skeletal system at multiple sites while solitary myeloma is a single focus of proliferation of plasma cells. Solitary myeloma can be present in bone or may have a soft tissue origin. Solitary myeloma of soft tissues is known as extramedullary plasmacytoma. Just as lymphomas or leukemias, multiple myeloma is a very common type of blood cancer with the incidence of one in every one hundred and fifty persons. But in contrast to this solitary myeloma and that too of soft tissue origin is a very rare clinical manifestation with an incidence of one in a million. Here is a case presentation of a soft tissue solitary myeloma or extramedullary plasmacytoma.

KEY WORDS: extramedullary plasmacytoma, myeloma, plasma cells

INTRODUCTION:
Extramedullary plasmacytomas are rare tumours of malignant origin. The term myeloma was first coined by Ruztky in 1873 and the earliest evidence of myeloma was found in Egypt. The first case of solitary myeloma was reported by Stout and Kennedy in 1949. Multiple myeloma is a plasma cell neoplasm involving skeletal system at multiple sites, while solitary myeloma in contrast to multiple myeloma is a single focus of proliferation of plasma cells. Solitary myeloma may involve a particular bone known as solitary myeloma of bone or may occur exclusively in soft tissues called as extramedullary plasmacytoma. Solitary myeloma of bone usually involves long and flat bones like vertebrae, spine or the bones of skull and its clinical features include pain and swelling in the region of the involved bones. Extramedullary plasmacytoma is a very rare clinical manifestation and amongst the few cases reported most of them were present in Nasopharynx, Nasal mucosa, gingiva and Subcutaneous tissues involving head and neck.

This disease usually manifests itself in the form of a localized soft tissue swelling which is extremely painful in nature. These plasmacytomas may invade the adjacent bones and metastasize to regional lymph nodes. Research studies show that if not diagnosed and treated at the right time, they may transform into multiple myeloma.1,2

CASE REPORT:
A 69 year old female patient reported to the Dental OPD with a complaint of swelling on the right side of the jaw. She noticed the swelling two months back. Initially, it was about the size of a lemon but then it suddenly increased in size and within a short period of two months, transformed into a massive swelling of 3x4 inches. The swelling was also accompanied by burning sensation over the affected area. Patient was not able to sleep due to throbbing pain and burning sensation. There were no complaints of bone pain, renal failure or any other systemic manifestations.

On examination it was found that a swelling was present on the right side of the mandible which extended from the angle of mandible to a line dropped to the angle of mouth posterolaterally, superiorly from a line joining the angle of mouth to the lobule of ear and upto the middle third of neck, inferiorly. The swelling was localized in nature, soft in consistency and showed no signs of inflammation. The regional lymph nodes were not enlarged.

On intraoral examination the patient was
found to be edentulous and interestingly, there were no signs showing intraoral origin of the swelling. On the basis of above mentioned complaints and examination a biopsy was performed and the results of the histopathological examination showed abundant amount of plasma cells. Sheaths of plasma cells were present with varying degree of differentiation along with a few lymphocytes and polymorphonuclear leukocytes – which is the characteristic feature of myeloma or plasmacytoma.

**DIAGNOSIS:**

Biopsy reports showed cells that are round or ovoid in shape with their nuclei eccentrically placed. Their nuclei showed clumping of the chromatin material exhibiting a typical cartwheel appearance and a perinuclear halo indicating the presence of Myeloma. Radiographic examination of skull and mandible did not reveal the presence of lytic or punched out lesions and there was no evidence of bone destruction. Lytic or punched out lesions are essentially present in multiple myeloma and solitary myeloma of bone, but absent in extramedullary plasmacytoma.

Patient was advised for plasma electrophoresis and the results of serum electrophoresis did not show hypergammaglobulinemia or presence of abnormal M proteins.
Hypergammaglobulinemia is always present in multiple myeloma, but absent in both the types solitary myeloma i.e. solitary myeloma of bone and extramedullary plasmacytoma. Excessive gammaglobulins produced by malignant plasma cells can easily be identified in serum electrophoresis and act as a vital key in the diagnosis of multiple myeloma and more importantly in the differential diagnosis of solitary myeloma from multiple myeloma.

Since the reports of serum electrophoresis and radiographic examination were normal and since there were no complaints of bone pain, renal failure, anemia or any other systemic manifestation the possibility of multiple myeloma was completely ruled out. However based on biopsy reports, it was diagnosed as a case of solitary myeloma and absence of radiographic findings confirmed it to be extramedullary plasmacytoma affecting mandibular and inframandibular region.

**DISCUSSION:**

Solitary myeloma remains undiagnosed in most of the cases and later on transforms into multiple myeloma. A case of extramedullary plasmacytoma of oesophagus has been reported in a 58 year old man. The chief complaints being dysphagia for solid foods. Two cases of extramedullary plasmacytoma of rectum and their treatment by radiotherapy have been reported. First patient was relieved in 36 months while the other in 3 months. Similarly another study analysed the clinical features and response to treatment of 35 patients with extramedullary plasmacytoma. Median age was 49 years.

**CONCLUSION:**

Solitary myeloma is a rare clinical manifestation with the incidence of 1 in a million. It can be found, during intensive surveillance, in significant quantum amongst the dental outpatients. However, many of these are missed due to lack of thorough clinical examination and tedious diagnostic requirements and finally emerge out as multiple myeloma. Virtually rare occurrence, exceptionally different presentation, intensively thorough course of investigation and probability of malignant transformation make solitary myeloma a crucial ailment.

**REFERENCES:**