Case Report

Solitary plasmacytoma: A case report

Sandya Rani Padma Guruvelli[1], Vandana Gangadharan[2], S Sri Krishna Prakash[3], Bhagyalakshmi Atla[4]

From [1]Post Graduate, [2]Professor, [4]Professor and Head, Department of Pathology, [3]Professor and Head, Department of ENT, NRI Institute of Medical Sciences, Visakhapatnam, Andhra Pradesh, India

ABSTRACT

Plasmacytoma is a discrete, unifocal, and monoclonal neoplastic proliferation of plasma cells. It may present as one of the three distinct clinical entities: multiple myeloma (MM), solitary plasmacytoma of the bone, and extramedullary plasmacytoma. Solitary plasmacytoma of the bone accounts for 3% of all plasma cell neoplasms. About 50% of the cases of solitary plasmacytoma of the bone transform into MM. The most commonly involved sites are vertebrae and long bones. It is extremely rare in jaws. We are presenting a case of solitary plasmacytoma of the maxilla in a 59-year-old female patient with comprehensive clinical, radiological, histological, and immunohistochemical features.

Key words: Contrast-enhanced computed tomography, Fine-needle aspiration cytology, Histopathology, Immunohistochemistry, Plasmacytoma

Plasmacytomas are a diverse group of lymphoid neoplasms characterized by clonal neoplastic proliferation of terminally differentiated B-lymphocytes (plasma cells or myeloma cells). The plasma cells were first described by Unna in 1891 and Schridde in 1905 [1]. Plasmacytomas are grouped under B-cell peripheral lymphomas according to the classification of the revised European-American International Lymphoma Study Group [1]. The incidence of plasma cell neoplasms is approximately 2.6–3.3/100,000 population [2]. The three distinct clinical entities of plasma cell neoplasms include multiple myeloma (MM), solitary plasmacytoma of the bone (SPB), and extramedullary plasmacytoma (EMP). MM is the most common of these neoplasms accounting for 65% of cases, which also represents 1% of all malignancies [2]. SPB and EMP are less common, localized forms that further evolve into disseminated MM within months or years after the initial diagnosis.

CASE REPORT

A 59-year-old female reported to the ear and nose throat outpatient department complaining of pain and swelling over the right cheek for 3 years. Past history revealed tooth extraction on the right side 3 years back. She complained of difficulty in opening the mouth and breathing from the right nostril. No details of past treatment were available with the patient.

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General examination revealed no abnormality and vitals were stable. Local examination showed a well-defined swelling measuring 10×7 cm extending from the right eye to the body of the mandible vertically and from the dorsum of the nose to the preauricular area horizontally (Fig. 1). The nose was deviated to the left side. There was no local rise in temperature. No palpable lymph nodes or signs of paresthesia were seen.

Oral examination revealed an intraoral extension of well-defined solitary erythematous growth measuring 3×3 cm in the right retromolar region at the site of previous tooth extraction. On palpation, the growth was soft to firm in consistency with a smooth surface and well-defined margins.

A contrast-enhanced computed tomography of facial bones was done which showed an enhancing exophytic mass lesion with multiple coarse calcifications and vascular channels most probably arising from the parapharyngeal space with destruction of the right orbit, ethmoid, turbinates, nasal septum, nasal cavity, hemi-maxilla, maxillary sinus, and zygomatic arch. Orthopantomogram showed the mouth to be deviated to the left side (Fig. 2).

Fine-needle aspiration cytology was performed from the swelling as per the standard technique and sent for cytological examination. Smears studied were cellular which showed sheets of discrete cells composed predominantly of plasma cells with eccentric nuclei and abundant cytoplasm along with lymphocytes and macrophages. Occasional binucleate plasma cells were also seen (Fig. 3). With these findings, a provisional diagnosis of

Correspondence to: Dr. Vandana Gangadharan, 02 Koyal, Naval Park, Visakhapatnam-530014, Andhra Pradesh, India. E-mail: vandanagangadharan@gmail.com

plasmacytoma was offered with an advice for biopsy. All other routine hematological and biochemical investigations were within normal limits. A complete skeletal survey showed no other bony lesions. Bone marrow aspiration was refused by the patient.

A core-needle biopsy of the swelling was done and sent for histopathological examination. Histopathological examination showed tumor cells arranged in diffuse sheets along with spheroidal eosinophilic acellular material (probably amyloid). Individual tumor cells were polygonal with abundant eosinophilic cytoplasm and eccentric nuclei. Foci of mild nuclear pleomorphism and a few foreign-body giant cells were seen (Fig. 4).

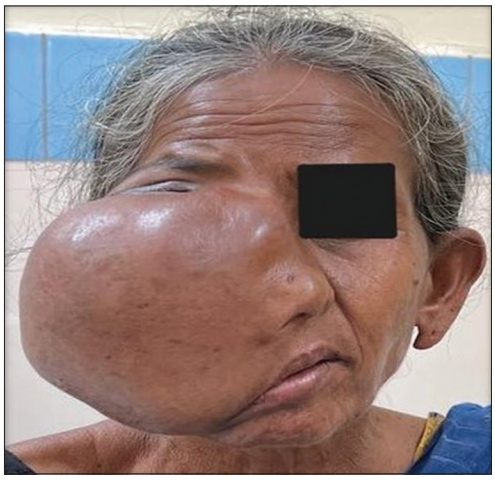

Figure 1: Clinical image of swelling over right cheek measuring 10×7 cm

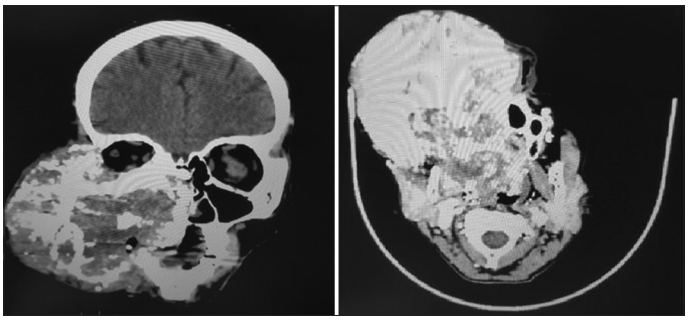

Figure 2: Contrast-enhanced computed tomography images of facial bones show exophytic mass with destruction of right orbit, ethmoid, turbinates, nasal septum, nasal cavity, maxilla, and maxillary sinus

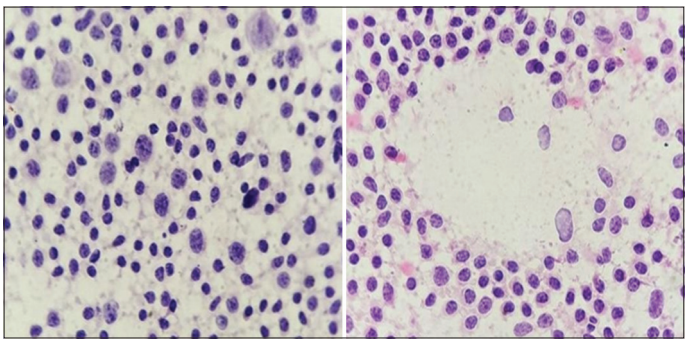

Figure 3: Fine-needle aspiration cytology smears show sheets of discrete cells composed predominantly of plasma cells, lymphocytes, and macrophages (Giemsa: 40X)

Considering these findings, the possibility of plasmacytoma was considered. Immunohistochemistry (IHC) was done for CD138, Kappa, and lambda. CD138 showed diffuse positivity. Kappa was positive in the tumor cells and lambda was negative (Fig. 5). In view of the clinical features, no evidence of end-organ damage with the absence of hypercalcemia, renal dysfunction, anemia, or bone disease (CRAB), histopathological findings, and IHC, a diagnosis of solitary plasmacytoma of maxilla was made. However, the patient refused any further intervention and was lost to follow-up.

DISCUSSION

Plasmacytoma is a lymphoid neoplastic proliferation of B cells that occurs in three forms – SPB, EMP in soft tissues, or MM. Solitary plasmacytoma of the bone accounts for 1–2% of all plasma cell neoplasms. It represents <5% of all plasma cell dyscrasias. The etiology of SPB remains uncertain but several hypotheses were proposed that implicated the role of radiation, chemical exposure, viruses, and genetic factors. Cytogenetic studies revealed loss in chromosome 13, 1p, 14q and gain in 19p, 9q, 1q, and interleukin-6, which is considered a principal growth factor in its pathogenesis [1].

The most common sites of SPB are long bones, and vertebrae [2]. Maxillary plasmacytoma is rare and its incidence increases with age. Solitary plasmacytoma of the bone occurs most frequently in patients between the ages of 50 and 80 years with a mean age of occurrence of about 60 years [3]. It is rare before the age of 40 [3]. It is more prevalent in men when compared to women, with the ratio being 2:1 [3]. The clinical presentation of solitary plasmacytoma of the maxilla is facial swelling, post-extraction bleeding, and sensory disturbances [4]. Our case was in harmony with the literature since the main complaint was swelling and pain, with a history of extraction but paresthesia was not reported.

The diagnosis of solitary plasmacytoma depends on the biopsy evidence of plasma cell proliferation and the absence of involvement of other bones [5]. There should be no evidence of organ damage including hypercalcemia, renal dysfunction, and anemia [6]. Confirmation of the presence of a monoclonal plasma cell population by IHC is mandatory. Radiology depicts osteolytic lesions which can be mono- or multilocular without any sign of the bone reaction [7,8].

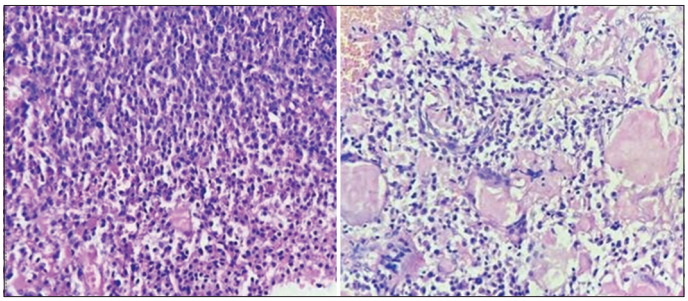

Figure 4: Photomicrographs showing sheets of plasmacytoid mononuclear cells with eosinophilic amyloid material. H&E 40X

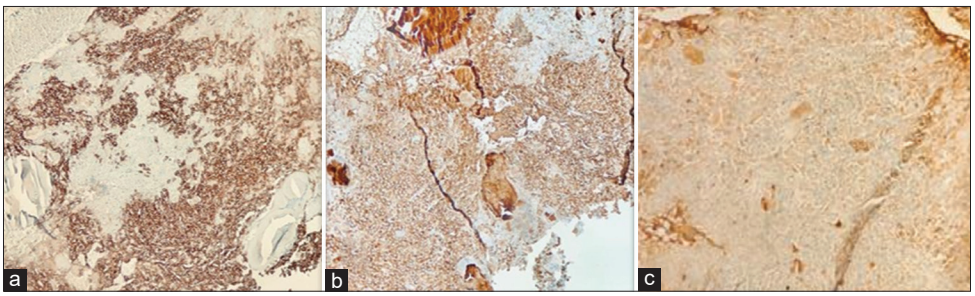

Figure 5: (a) Tumor cells show diffuse positivity for CD-138. Immunohistochemistry (IHC) ×40; (b) Tumor cells show diffuse positivity for Kappa. IHC ×40; (c) Tumor cell show lambda negative. IHC-40X

In many instances, plasmacytoma may be misdiagnosed as a benign lesion such as plasmacytosis [9]. These can be clinically differentiated based on tumor size, bone involvement, evolution, and most importantly, histology which is conclusive with IHC as an adjunct. A plasmacytosis is plasma cell granuloma which is polyclonal and not neoplastic. In contrast, a plasmacytoma is monoclonal and has a single kappa or lambda light chain.

There have been no therapeutic advances in the treatment of solitary plasmacytoma over the years. Treatment methods include local surgery (curettage of the lesion), local irradiation, systemic chemotherapy, or a combination of these methods. Radiation is the mainstay of therapy for plasmacytoma, as it is highly radiosensitive [6,10]. Surgical excision is reserved for cases where there is a loss of anatomic structural integrity or emergent decompression.

Despite excellent local control rates, the majority of patients with solitary plasmacytoma eventually progress to MM. Studies have shown a 5-year overall survival rate of 70% and a 5-year disease-free survival rate of 46%, with the median time to develop of MM being 21 months, with a 5-year probability of 51% [11]. Therefore, it is important to first assess patients for a complete response after radiation therapy and follow them to detect possible recurrence. This is done with laboratory evaluation that would include a complete blood count, complete metabolic panel, lactate dehydrogenase, beta-2-microglobulin, urine protein electrophoresis, serum protein electrophoresis, free light chains, and serum quantitative immunoglobulins. Laboratory evaluation should be performed roughly every 3 months for the first 2 years following radiation treatment and then every 6 months [10,11]. We advised post-surgical radiotherapy and chemotherapy, but the patient was lost to follow-up.

CONCLUSION

SPB is a rare entity and even rarer is its presentation in the maxilla. The diagnosis requires histological and immunohistochemical evidence, and proving the solitary character, once the diagnosis is made. The management must be rapid, with careful monitoring, because solitary plasmacytoma of the bone, despite its localized form, has a higher propensity to transform into MM.

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