Original Research Paper

Volume - 12 | Issue - 06 | June - 2022 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Otorhinolaryngology

SOLITARY BONE PLASMACYTOMA OF THE LEFT ZYGOMA REGION: A RARE CASE REPORT

Sagung Rai Indrasari	Department of Otorhinolaryngology Head and Neck Surgery, Dr. Sardjito Hospital, Yogyakarta, Indonesia
Camelia Herdini	Department of Otorhinolaryngology Head and Neck Surgery, Dr. Sardjito Hospital, Yogyakarta, Indonesia
Danu Yudistira	Department of Otorhinolaryngology Head and Neck Surgery, Dr. Sardjito Hospital, Yogyakarta, Indonesia
Anisa H Khoiria*	Universitas Gadjah Mada, Yogyakarta, Indonesia*Corresponding Author
Yosephine Nina	Universitas Gadjah Mada, Yogyakarta, Indonesia

ABSTRACT **Background:** Solitary Bone Plasmacytoma (SBP) is a characterized by a mass composed of monoclonal plasma cells in bone without any proof of systemic disease attributing to myeloma. It is a rare form of plasma cells in bone that represents only 5 – 10% of all plasma cell neoplasm. The diagnosis requires solitary bone lesion confirmed by skeletal survey, plasma cell infiltration proven by biopsy, normal bone marrow biopsy, and no myeloma-related organ failure. **Purpose:** Reporting one rare case of solitary bone plasmacytoma of the left zygoma region treated with surgical excision followed by radiotherapy. **Case Report:** A 61 years old male came with complains of swelling in the left cheek for 2 years. Histopathological examination by fine needle aspiration biopsy of the left zygoma region showed atypical plasmacytoid cell with suspected plasmacytoma which then confirmed by positive CD 138 staining. Clinical Question: What is the management of solitary bone plasmacytoma of the zygoma region? **Review Method:** Searching for literature evidence through Google Scholar. **Result:** The search obtained 11 journals that met the inclusion and exclusion criteria. There were similarities in clinical symptoms and management with this reported case. **Discussion:** In the reported case, the clinical features of SBP are swelling or mass in the zygoma bone spreading to maxillary bone with intermittent mild pain with no ear, nose, or throat complain, and no myeloma related organ dysfunction. The diagnosis was determined by history taking, physical examination, and supporting examination by fine needle aspiration biopsy which was confirmed by the pathology anatomy feature of the post-surgical specimen and CT imaging. **Conclusion:** Solitary bone plasmacytoma manifests itself as a single osteolytic lesion without plasma cytosis of bone marrow and constitutes approximately 5% of all plasma cell neoplasms. SBP reinforces for the meticulous diagnostic work up. In this case, surgical excision followed radiation is the treatment of choice, as plasma cell neoplasm are highly radiosensitive.

KEYWORDS : Plasmacytoma, Solitary Bone Plasmacytoma, Surgical Management, Radiotherapy

INTRODUCTION

Solitary plasmacytoma of bone is a rare neoplasm that occurs less than 5% of all plasma cell neoplasm. [1] It is a form of a rare plasma cell dyscrasia, which were subclassified as solitary bone plasmacytoma (SBP) or solitary extramedullary plasmacytoma (SEP) depending on the site of involvement, whether it invades the bone or soft tissue. [1][2]

The diagnostic criteria for this disorder were as follows: a solitary lesion of the bone due to biopsy proven clonal plasma cells, bone marrow not consistent with multiple myeloma, normal skeletal survey, no anemia, no hypercalcemia, and no renal impairment that correspond to plasma cell disorder. [1][2] The diagnosis requires biopsy that proves an infiltration of clonal plasma cells into a single lesion. [2] SBP most frequently occurs in the axial skeleton such as vertebra, while SEP is generally observed in head and neck, but these two kinds of plasmacytoma have quite difference in clinical features and prognosis. [2]

Because of its rarity, most studies were based on relatively small number of patients and limited possibility to reach any definitive conclusion. In the previous study, the spine and upper airway were the most common site involved in SBP with the percentage of 48% and 50% for SEP. [3] Solitary bone plasmacytoma mostly occurs in the axial skeleton and is always associated with bone pain, pathologic fractures, or nerve involvement. [3][4]

The therapeutic approaches to SBP mainly include radiotherapy, surgery, and chemotherapy. [3][4] Surgery is a milestone for the histological diagnosis in the form of biopsy or deletion of the lesion both partial and total, also considered as specific treatment for plasmacytoma with particular localization. [3][4] Radiation therapy of at least 40 Gy in limited fields are recommended for the management of SBP. Although some studies mention that radiotherapy is associated with good survival, surgery should be required if the pathologic features are present in the involved bone, when immediate decompensation of a tumor is necessary, or when moderate doses of radiotherapy are problematic due to the presence of surrounding critical organs. [4] However, the function of chemotherapy remains unclear, there is no consensus on chemotherapy use for patient with SBP. [4]

While it is difficult to identify the prognostic factors due to the homogenously distributed data from the previous cases, different factors such as age, lesion size, localization (bone or soft tissue), presence of M-protein that could be a sign of multiple myeloma transformation have been reported. [4] Attention should be paid to local bone involvement in routine clinical work, especially patients with risk factors for multiple myeloma require close follow up, more investigations to show how the progression into multiple myeloma could be prevented or delayed. [4]

The purpose of presenting this article was to report one rare case of solitary bone plasmacytoma in order to provide new insight of the disease and information regarding clinical features, management, and promoting early diagnosis to avoid the disease progression into multiple myeloma.

CASE STUDY

A 61 years old male patient presented with swelling in the left cheek for 2 years, the patient felt that the swelling was rapidly grow in the last 6 months (Figure 1). The swelling from the mass causing intermittent pain, fullness on the cheek, and discomfort. The patient denied any nasal, eye, ear, and throat symptom. The physical examination showed that ear, nose, and throat were within normal limit. Nasal endoscopy showed no mass, no thrusting from the mass to the nasal cavity. Facial examination revealed asymmetric appearance of the face, with swelling at the left zygoma region. From the palpation, the size of the mass was about 5x5x3 cm, no tenderness or any sign of inflammation, the consistency was hard and fixated to the zygoma bone expanded to the left maxillary region. No palpable neck mass was found. The patient had been working as a welder for the past 20 years with prolonged exposure of chemical and asbestos dust to the face.

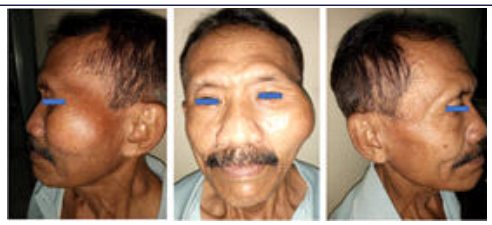

Figure 1. Clinical appearance before surgery

The punch biopsy could not be done due to the accessible protruding mass so we decided to do fine needle aspiration biopsy (FNAB). Histopathology examination of the FNAB revealed atypical plasmacytoid cell with suspected plasmacytoma or chronic suppurative inflammation, further immunohistochemistry staining with CD 138 was needed to confirm the diagnosis. We proceed the CD 138 staining and received positive result that indicate the mass of the left zygoma region was plasmacytoma (Figure 2).

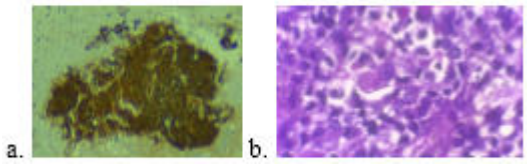

Figure 2. a). Histopahology examination CD 138 staining, b). Histopathology of the tumor excision confirmed plasmacytoma.

We also did CT imaging with contrast agent and discover that the mass was originated from bone with malignant feature at the lateral aspect of the left zygoma bone (Figure 3). No abnormality found in paranasal sinuses and both nasal cavities, no cervical lymphadenopathy was detected. Further diagnostic analysis was laboratory assessment of complete blood count, serum calcium, and renal function. All tests yield normal results. Skeletal survey showed no other bone lesion except in left zygoma region was detected. Chest X-ray also showed normal appearance.

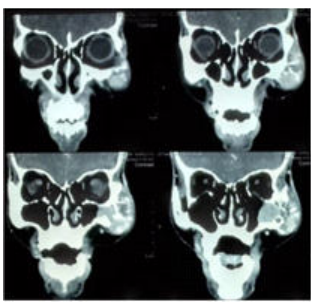

Figure 3. CT imaging of the facial bone

Based on the history taking, physical examination, histopathology, laboratory, and imaging, we diagnosed the patient with solitary bone plasmacytoma of the left zygoma region, therefore we planned to do a surgical excision of the mass followed by radiotherapy.

Surgical procedure using subcilliar approach was done to remove the mass. the incision is placed closed to the lid, extended laterally across the left zygoma region following the skin tension line, continued with blunt dissection until the tumor margin was identified (Figure 4a). The tumor was hard in consistency, fixated and embedded in the left zygoma bone, extending to the lateral part of the maxillary bone with minimal destruction. The tumor was then dissected bluntly and sharply from the adjacent tissue. The base of the tumor was connected to the lateral part of the left maxillary bone (Figure 4b).

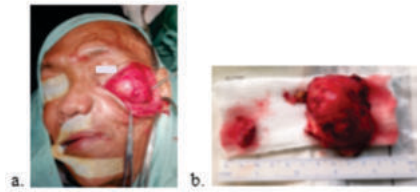

Figure 4. a) Surgical incision, b) Tumor specimen from the surgery

Therefore, we decided to also resect the left maxillary bone that was embedded by the tumor, by using drill we managed to take out the tumor and its capsule that embedded in the maxillary bone. The dimension of the tumor was 5 x 4.6 x 2.4 cm. The resected tumor and bone were then sent for histopathologic examination to confirm the previous FNAB result. The surgical wound was sutured layer by layer. The wound was dressed every 24 hours with administration of antibiotic, anti-inflammatory, and analgesic injection. The patient was discharged from the hospital 3 days after surgery and advised to do follow up to the outpatient clinic 4 days later.

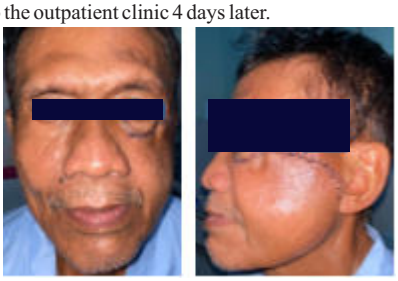

Figure 5. Clinical appearance 7 days after surgery

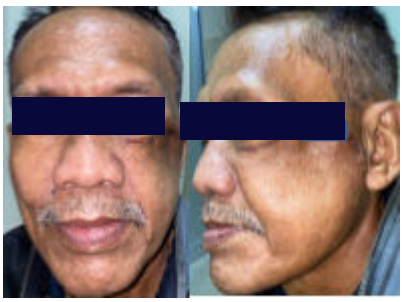

Figure 6. Clinical appearance 2 weeks after surgery

The first follow up, the surgical wound was in good condition. The stiches were removed partially 7 days after surgery at the first follow up (Figure 5). Then removed completely at the second follow up, 14 days after surgery (Figure 6). The patient then referred to the cancer institute for complete treatment which include radiation therapy.

CLINICAL QUESTION

What is the management of solitary bone plasmacytoma of the zygoma region?

REVIEW METHOD

Literature search was carried on in December 2021 using keywords "solitary bone plasmacytoma" AND "diagnosis" AND "management" NOT "multiple myeloma" through Google Scholar. Selection was based on inclusion criteria as follows: 1) Head and neck region, 2) Contained the diagnosis steps and management, 3) Managed by surgery followed by radiation therapy. Exclusion criteria were 1) Progression into multiple myeloma and 2) Multiple regions plasmacytoma.

RESULT

The result of literature search through Google Scholar, obtained 20 articles published in the last 10 years, selection was based on inclusion and exclusion criteria, we found 11 articles which were relevant with the topic.

DISCUSSION

Solitary bone plasmacytoma appeared as a localized accumulation of neoplastic monoclonal plasma cells without any proof of a systemic plasma cell proliferation disorder that correlates with myeloma. It is a very rare form of plasma cell neoplasm, represent 5-10% of all plasma cell neoplasms. [5] Plasmacytoma was classified into 2 groups based on its location, solitary bone plasmacytoma (SBP), which mostly occur in bones of the axial skeleton such as vertebrae and skull, also extramedullary plasmacytoma (EMP) which mostly observed in head and neck, frequently in nasal cavity and nasopharynx. The median age for either SBP or EMP is 55 years old, with male-to-female ratio is 2:1. Incidence rate rises exponentially by increasing age; but less prominent at older ages in comparison with multiple myeloma. [5]

Solitary bone plasmacytoma is an immunoproliferative monoclonal

disease, accounting for 3-7% of all cases of plasmacytoma. Although the etiology remains uncertain, certain etiologic agents such as radiation, exposure to chemicals, viruses, and genetic factors have been implicated. Cytogenic studies in plasmacytoma reveal loss in chromosome 13, 1p, 14q, gain in 19p, 9q, 1q, and IL-6 is considered to be the principal growth factor. Radiologically the lesion is osteolytic with unilocular/multilocular radiolucency. Osteoclastic activation by cytokines increases the osteoclast numbers in areas invade by malignant plasma cells, resulting in an osteolytic lesion. [5][6]

Diagnostic analysis consists of history, physical examination, complete blood count, bone marrow biopsy, serum protein electrophoresis, evaluation of the urine for myeloma protein, and skeletal survey. Staging system for solitary bone plasmacytoma is regarded as stage I myeloma and contains all of the following criteria: hemoglobin < 10 g/dL, normal level of serum calcium, normal bone structure or solitary plasmacytoma only, and low M-component (IgG < 5 g/dL, IgA < 3g/dL, urine light chains < 4g/24 h) Clinically, patient can present with bone pain, localized swelling of the area with pressure symptoms on the surrounding structure. [5][6]

Skeletal survey is beneficial to detect osteoblastic response to bone destruction. CT imaging may be more helpful to detect the extend of bone destruction. Plasmacytoma is very difficult to confirm the diagnosis without radiological, histopathological, immunohistochemical, and other supportive investigative findings from available modalities. In our case along with radiological, histopathological findings, support of immunohistochemistry positivity for CD 138 was also done to achieve the right diagnosis. [5][6][7] Currently, the standard care for SBP is definitely radiotherapy. SBP is a highly radiosensitive disease with excellent local control rate of > 80%. In some cases, surgical intervention may be required for bone instability or for rapidly progressive neurological symptoms like spinal cord compression. For optimal treatment with sufficient local control, a moderate dose RT combined with surgery is occasionally suggested. RT should be used after gross total excision to eradicate microscopic residual disease. Even though the optimal dose of RT has not yet been established for SBP, it is recommended that a radiation dose of at least 40Gy in four weeks is necessary to obtain local control. [5][6][7][8] Adjuvant chemotherapy has no beneficial effect on disease control or prevention of progression to multiple myeloma. [7][8][9]

There are three patterns of failure which includes development of multiple myeloma, local recurrence, and development of new bone lesions without MM. [8][9] SBP has a significantly higher risk for progression to myeloma at a rate of 65-84% in 10 years and 65-100% in 15 years. Lesion size of minimum 5 cm, age 40 years and over, spine lesions, RT dose, high M protein levels, existence of light chains, and persistent M protein after treatment can influence the outcome, it may also indicate the presence of higher risk of progression to MM. [8][9]

Treatment response was evaluated to the response evaluation criteria in solid tumors (RECIST). Partial response was defined as reduction of tumor size by at least 30% in the imaging studies and disappearance of serum M-protein. [9][10] Following completion of radiotherapy, patients visited hospital every 3 months for the first two years, every 6 months for the next 3 years, and annually thereafter. Follow up studies consists of CBC, blood chemistry, serum and urine assays to detect M-Protein, and imaging to assess the treated area. [9][10][11]

Our reported case showed how difficult it is to establish the diagnosis of SBP, with regards to common swelling symptoms without any ear, nose, or throat complained, aggravated by difficulty in obtaining tissue sample. The etiology is uncertain, many factors may be contributed to the development of this disease, particularly in this case, there is an occupational exposure to certain chemical that can relate to the probable etiology. The case is very rare for sure because from the literature, this disease mostly affects axial skeleton, rarely in the facial bone, for that matter, physician should always be alert of the occurrence of solitary bone plasmacytoma of the head and neck region.

REFERENCES:

1. Warsame R, Gertz MA, Lacy MQ, Kyle RA, Buadi F, Dingli D, et al. Trends and outcomes of modern staging of solitary plasmacytoma of bone. Am J Hematol. 2012; 87: 647-51.
2. Pham A, Mahindra A. Solitary Plasmacytoma: a Review of Diagnosis and Management. Curr Hematol Malig Rep. 2019; 14: 63-9.
3. Finsinger P, Grammatico S, Chisini M, Piciocchi A, Foa R, Petrucci MT. Clinical features and prognostic factors in solitary plasmacytoma. British Journal of Haematology. 2016; 172: 554-60.
4. Shan-qi G, Le Z, Ya-fei W, Bao-cun S, Lian-Yu Z, Jin Z, et al. Prognostic factors associated with solitary plasmacytoma. Onco Targets and Therapy. 2013; 6: 1659-66.
5. Kilciksiz S, Celik OK, Agaoglu FY, Haydaroglu A. A Review for Solitary Plasmacytoma of Bone and Extramedullary Plasmacytoma. The Scientific World Journal.2011; 2012 : 1-6.
6. Rao K, Priya NS, Umadevi HS, Smitha T, Reshma V, Brace SA. Solitary Bone Plasmacytoma of the Maxilla – A rare case report. Journal of Clinical Dental Science. 2011; 2(1): 37-40.
7. Kamal M, Kaur P, Gupta R, Gupta S, Singh S. Mandibular Plasmacytoma of Jaw – A Case Report. Journal of Clinical and Diagnostic Research. 2014;8(8): 20-21.
8. Sasaki R, Yasuda K, Abe A, Uchida N, Kawashima M, Uno T, et al. Multi-institutional Analysis of Solitary Extramedullary Plasmacytoma of the Head and Neck Treated with Curative Radiotherapy. I J Radiation Oncology Biol. Phys. 2012; 82(2): 626-34.
9. Suh YG, Suh CO, Kim JS, Kim SJ, Pyun HO, Cho J. Radiotherapy for solitary plasmacytoma of bone and soft tissue, outcome and prognostic factors. Ann Hematol. 2012; 91: 1785-93.
10. Li QW, Niu SQ, Wang HY, Wen G, Li YY, Xia YF, Zhang YJ.Radiotherapy Alone is Associated with Improved Outcomes Over Surgery in the Management of Solitary Plasmacytoma. Asian Pac J Cancer Prev. 2015; 16(9): 3741-5.
11. Elsayad K, Oertel M, Konig L, Huske S, Le Ray E, Meheissen MAM, et al. Maximizing the Clinical Benefit of Radiotherapy in Solitary Plasmacytoma: An International Multicenter Analysis. Cancers. 2020; 12(676): 1-12.