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CASE REPORT

Solitary extraosseous plasmacytoma

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Key Clinical Message

Plasma cell neoplasms are characterized by a neoplastic plasma cell lineage which produces a monoclonal immunoglobulin. These neoplasms can present as a single lesion (solitary plasmacytoma) or as multiple lesions (multiple myeloma). Solitary plasmacytomas most frequently occur in bone (plasmacytomas of bone), but can also be found outside bone in soft tissues (extramedullary plasmacytomas).

Keywords

Extramedullary, multiple myeloma, plasmacytoma, solitary extramedullary plasmacytomas.

Introduction

We do not understand yet why some patients develop multiple myeloma and others a single plasmacytoma [1, 2], but that might be related to differences in the chemokine receptor expression profiles of the malignant plasma cells or cellular adhesion molecules [3]. Solitary extramedullary plasmacytomas (SEP) are extremely rare tumors that arise outside of the bone marrow in the absence of any sign of multiple myeloma [4, 5]. They are solitary lesions, and are often seen in the head and neck regions [6, 7], mainly in the upper aerodigestive and to a lesser extent in the gastrointestinal tract (GI) tract, bladder, central nervous system (CNS), thyroid, breast, testes, parotid gland, lymph nodes, as well as in skin.

Solitary extramedullary plasmacytomas tends to occur during the fifth and seventh decades of life, rarely in younger population [8].

Extramedullary plasmacytomas (EMPs) can arise in patients with multiple myeloma at any time during the course of the disease and in one-third of the cases, resulting in a worse clinical outcome that should not be confused with SEP [9, 10].

Case Presentation

This particular case involves a 40-year-old Hispanic male with past medical history of mental retardation, anxiety, and dyslipidemia who lives in a group home. Patient presented initially to his Primary care physician (PCP) for an annual physical. During routine examination, a neck mass was identified on the base of the lateral right side of the neck anterior of the sternocleidomastoid that measured 3 cm by 3 cm nontender with slight mobility. He denied any constitutional symptoms such as fever, chills, sweating, weight loss or change in diet or bowel habit, easy bruising, or hoarseness.

Patient was referred to ENT, and neck soft tissue CT scan with and without contrast was performed which showed enhancing 3.4 × 2.8 × 5.2 cm mass right retromandibular region just anterior to the sternocleidomastoid muscle with mass effect pushing the carotid vessels posteriorly with adjacent bony destruction; metastasis was not excluded by this test. Also, the scan was showing anterior adjacent mass approximately 0.8 × 1.1 cm that may present metastasis or mildly enlarged lymph node.

Upon seeing ENT doctor, Fine needle aspiration (FNA) was done on the site and referred to a medical oncologist

for this suspicious mass indicating the malignant pathology. FNA result was not diagnostic as it showed cuboidal to columnar histologically benign appearing cells along with many small to medium size lymphocytes with differential diagnosis of: (i) salivary gland neoplasm, (ii) branchial cleft cyst, or (iii) possibility lymphoid proliferative disorder. Based on the FNA result, lymphoma could not be ruled out and surgical biopsy was recommended at that time. Initial blood work up was done showing monocyte was elevated of 11.3%.

Blood work up:

Cr	0.9
WBC	5.9
Hb	15.5
Hct	45.3
MCV	90.5
Plt	283
Granulocytes	62.8%
Lymphocytes	28.2%
Monocytes	8.0%
Glucose	87
Na	142
K	4.3
AST	19
ALT	27
AlkPhos	80

Initial differential diagnoses by ENT physician involved Castleman disease versus plasma cell neoplasia. The report came back favoring plasma cell neoplasm "possibility of primary lymph node plasmacytoma by systemic involvement exclusion."

Open biopsy was done with frozen section and it was found to have an extensive plasma cells with no carcinoma but some cells showing binucleated forms and atypical nuclei with final diagnosis of primary lymph node plasmacytoma (Figs 1 and 2).

There was kappa light chain restriction by Immunohistochemistry (IHC).

The following observations were made:

CD138 staining: positive flow cytometry: nondiagnostic.
Beta-2 microglobulin: normal range with no elevation immunofixation.
Quantitative immunoglobulins IgG/A/M panel: normal.
Serum K/L light chains ratio: normal.
PET CT scan showing mildly enlarged right level 2B lymph nodes measuring 14 mm with SUV of 4.1 and few adjacent smaller level 2B lymph nodes which are a normal size with low-grade metabolic activity.
BM Bx was done and showed no myeloma, norm cellular marrow with maturing trilineage.
Hematopoiesis – no definitive morphologic or immunephenotypic evidence of clonal expansion of plasma cells or involvement by a mature B-cell non-Hodgkin lymphoma – mildly increased iron stores
Kappa and lambda: stain a few polytypical plasma cells in normal number and distribution (Figs 3–6).

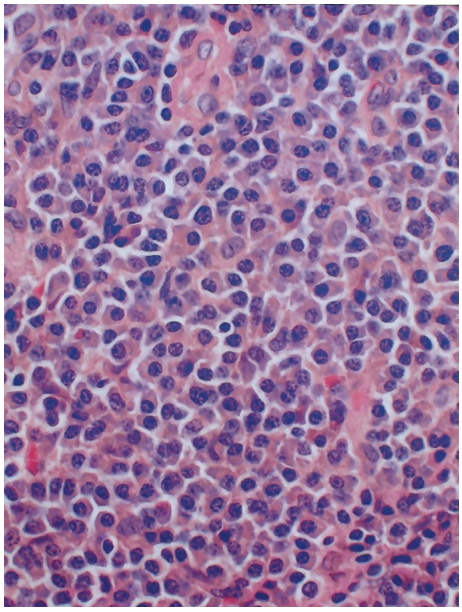

Figure 1. Plasmacytoma microscopic image.

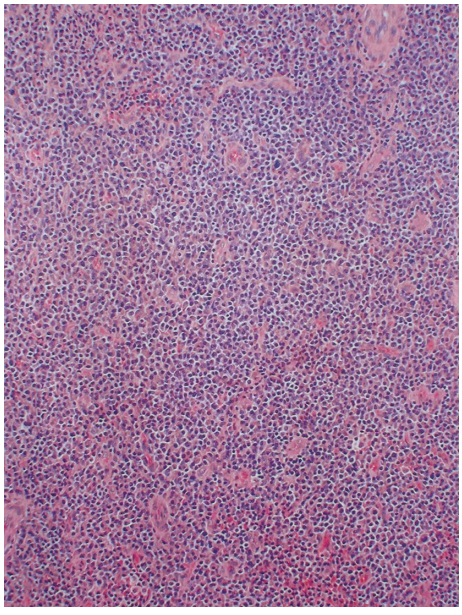

Figure 2. Plasmacytoma microscopic image low power view.

Initial recommendation by the oncologist was radiation therapy for definitive treatment for localized plasmacytoma of the neck which should likely provide adequate disease control on-site.

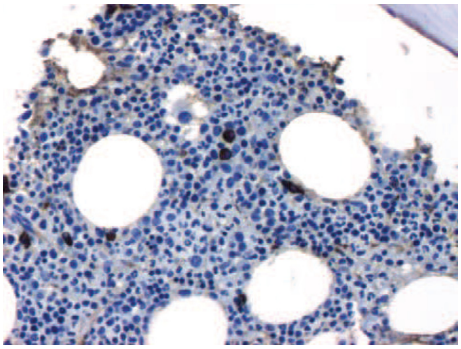

Figure 3. Kappa stain.

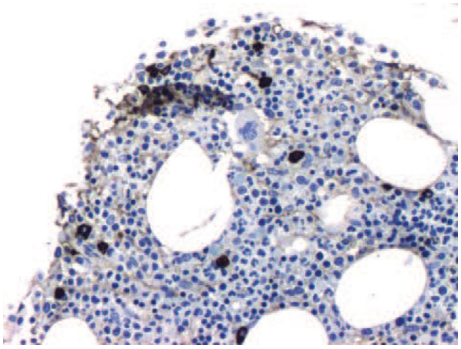

Figure 4. Lambda stain.

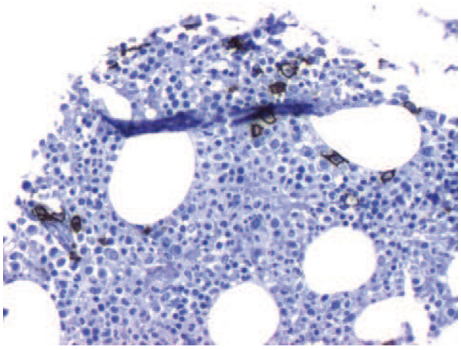

Figure 5. CD 138 stain.

Patient received 4500 cGy in 25 fractions of radiation treatments on the right neck with 3060 cGy in 17 fractions. The primary tumor site received additional 1440 cGy in eight fractions of the total of 4500 cGy in 25 fractions.

After radiation, patient was observed and monitored periodically and clinically did not show any evidence of residual disease other than superficial skin changes from the radiation which resolved later.

The patient will be at risk for developing plasmacytoma at other locations, hence he will be under close follow up.

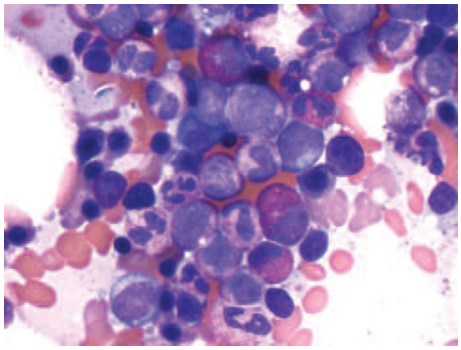

Figure 6. Aspirate smear stain.

Discussion

In this report, we present a case of SEP with no involvement of the bony structure or bone marrow. The case is quite rare as it represents 2–10% of all the multiple myeloma cases [11, 12]. Solitary extra osseous plasmacytoma has a unique characteristic of being undetectable disease elsewhere on PET/CAT scan.

A plasmacytoma is a unique solitary mass of neoplastic monoclonal plasma cells in either bone or soft tissue (extramedullary).

Plasmacytoma has been classified into three subtypes [13]. The most common type is multiple myeloma, which is usually a disseminated disease and characterized by abnormal M protein. The other two types, solitary plasmacytoma of the bone and EMP of the soft tissue, are considerably less common. EMPs present in <5% of plasma cell neoplasms and often (>80%) originate in the head and neck region [16]. Anatomically, we can divide the SEP into two groups: (i) plasmacytoma of the skeletal system (SBP) and (ii) EMP [21, 24].

The diagnosis of EMP of the soft tissue has been based on the following criteria: (i) pathological tissue evidence of monoclonal plasma cells involving a single extramedullary site; (ii) no bone marrow involvement; (iii) no anemia, hypercalcemia or renal impairment caused by plasma cell dyscrasias; (iv) negative skeletal survey results; and (v) low serum or urinary levels of monoclonal immunoglobulin [2]. The etiology of this disease remains unknown, but factors, such as chronic irritation from inhaled irritants or viral pathogenesis, have been previously indicated [17, 18]. Radiotherapy remains the mainstay for the management of EMP. As it considered radiosensitive, with a local control rate of 90–100% with a rate of conversion into MM is 31%. Moderate dose RT of at least 40 Gy using limited radiation fields is recommended [14, 15, 17, 19, 20]. Because of the high rate of recurrence and progression to multiple myeloma, follow-up radiological and electrophoresis assessment is required

following treatment [22]. The overall 10-year survival rate is ~70% [2, 16, 23]. A literature search revealed no publications supporting the use of surgery alone to treat EMP [24].

Conflict of Interest

None declared.

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