



Case Report

Cytodiagnosis of extramedullary plasmacytoma

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ABSTRACT

Extramedullary plasmacytoma is a rare malignancy constituting 3-5% of all plasma cell malignancies. Nearly 80% occurs in the upper autodigestive tract, while other sites include gastrointestinal tract, lymph node, bladder, CNS, breast, thyroid, testis, parotid and skin.

The utility of FNAC in the diagnosis of plasmacytoma has only been elaborated in few cases to date. A 50 years old man presented to our hospital with a mass on the chest wall measuring 3x2x2cms for one month. A cytological diagnosis of plasmacytoma was made which was confirmed on histopathology.

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1. Introduction

Extramedullary plasmacytoma is a rare plasma cell malignancy with a myriad of varying presentations, from being the sole manifestation, to a metastatic lesion from an extra medullary plasmacytoma present elsewhere, in the bone as a solitary plasmacytoma or as part of the disease process of multiple myeloma.

The sites of presentation vary widely, and have to be distinguished from other malignancies, infections and chloroma.¹

Plasma cell malignancies form a continuing spectrum of diseases, of which Plasma Cell Myeloma, solitary Plasmacytoma & the Extra medullary plasmacytoma form the integral components; the latter being rare.²

A mass of malignant plasma cells occur in extra medullary plasmacytoma; of which 5% are osseous and the remainder are extraosseous.

The most common extra osseous sites of development include oropharynx & nasopharynx while a minority involved draining lymph nodes. No evidence of increased plasma cells at other bone marrow sites, additional lesions on skeletal survey or fulfillment of clinical criteria for plasma cell myeloma are noted. A younger age of

presentation is noted when compared to overt myeloma; although eventual development of plasma cell myeloma is observed in 15% of extra osseous plasmacytoma & 2/3rd of patients with osseous lesions.³

2. Case History

A 50 years old man presented with mass on the chest wall measuring 3x2x2cms for one month. Cytological smear obtained by FNAC revealed groups & clusters of plasma cells in different stages of maturity in a highly cellular smear. The atypical plasma cells had increased N:C ratio, eccentric nuclei with prominent nucleoli and fine granular cytoplasm. Binucleate plasma cells & atypical mitotic figures were also noted. A cytologic diagnosis of plasmacytoma was made (Figure 1). The patient underwent an excision biopsy, which revealed sheets of plasmacytoid cells, with eccentric nuclei, conspicuous nucleoli & abundant eosinophilic granular cytoplasm. [Figure 2] Immunochemistry studies revealed a positivity for CD138 & negativity for CD20 & CD3, reiterating the diagnosis. The patient underwent a bone marrow examination to rule out marrow involvement, which showed none. A final diagnosis of Extramedullary plasmacytoma was made; following which the patient was started on therapy and is presently responding well to treatment.

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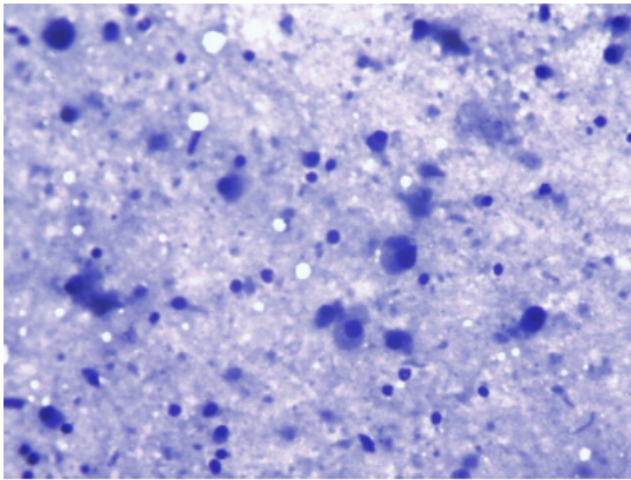


Fig. 1: Highly cellular smear composed of clusters of plasma cells with eccentric nuclei prominent nucleoli and finely granular cytoplasm. [Giemsa stain 400x]

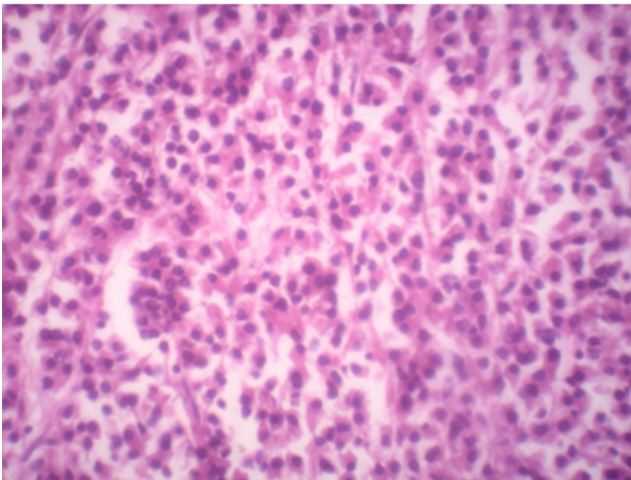


Fig. 2: Sheets of plasmacytoid cells with eccentric nuclei and abundant eosinophilic granular cytoplasm. [H&E Stain 400x.]

3. Discussion

Extra medullary plasmacytoma are rare neoplasms; often the initial manifestation of plasma cell myeloma. Presently there is limited data regarding the diagnosis, staging and natural history of the disease.⁴

Based on the location, solitary plasmacytoma maybe categorized as Plasmacytoma of the skeletal system(SBP) & Extramedullary plasmacytoma(EMP). Criteria for diagnosis of extra medullary plasmacytoma are the following:

Tissue biopsy revealing tumor cells with monoclonal plasma cell histology.

Plasma cell infiltration of bone marrow should not exceed 5% of all nucleated cells.

Serum M component, if present must be low.

No evidence of osteolytic bone lesions, other tissue involvement, hypercalcemia or renal failure.

The origin of a plasmacytoma can be from anywhere in the body. Plasma cells in the bone marrow give rise to a solitary bone plasmacytoma (SBP), while those in mucosal surfaces are the origin of extramedullary plasmacytoma (EMP).

Chromosomal analyses reveal recurrent losses of chromosome 13,1p, 14q and gains of 19p, 9q & 1q. A key role of interleukin-6(IL-6) has been noted in plasma cell disorders.

The occurrences of solitary bone plasmacytoma & extra medullary plasmacytoma are <5% & 3% of plasma cell malignancies respectively.

While the overall survival rate of extra medullary plasmacytoma is 70%, progression rate to plasma cell myeloma stands at around 11-30% within 10 years of diagnosis.

The median age at diagnosis is 55 years.⁵

While the site of presentation can vary widely, the most common(80-90%) site is the head and neck region, particularly the aerodigestive tract. The paranasal sinuses, pharynx, nasal cavity, oral mucosa & gums are seen to be involved in 80% cases. The most common manifestation is a mass that invades or compresses the surrounding structures.⁶ Local nodes are involved in 30-40%, either at presentation or at relapse.⁷

Soft tissue plasmacytoma was classified by Wiltshaw into three clinical stages:

Stage 1- Extramedullary site only.

Stage II-Regional lymph node involvement.

Stage III- Metastasis to multiple sites; no longer a solitary plasmacytoma.

Currently the approved treatment is radiotherapy. A 6 weekly periodic evaluation for progression to plasma cell myeloma is recommended for the initial 6 months for both solitary bone plasmacytoma & extra medullary plasmacytoma. In 2-3 years following diagnosis, distant metastases are seen to occur; hence a close follow up during this period is recommended.⁸

4. Conclusion

The rapid & accurate diagnosis of plasmacytoma by FNAC allowed for early initiation of treatment; thus this case highlights the diagnostic utility of this simple tool as part of the workup of a case; and ultimately therapeutic benefit for the patient.

5. Source of Funding

None.

6. Conflict of Interest

None.

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