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CASE REPORT

Extramedullary Plasmacytoma of Soft Tissue Neck

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ABSTRACT

Plasmacytoma is an extremely rare and discrete solitary mass of neoplastic monoclonal plasma cells. Extramedullary plasmacytoma tends to occur during the fifth and seventh decades of life and is rarely diagnosed in younger patients. In this study, a 55-year-old patient presented to us with complaint of changes in voice, and difficulty in swallowing and breathing. On examination, there was a retropharyngeal mass which is extending to lateral pharyngeal wall and toward nasopharynx. Contrast computed tomography scan shows soft tissue density involving retropharyngeal space and involving nasopharynx and left parapharyngeal space. For this, the patient underwent excision of this mass under General Anaesthesia (GA) biopsy report was suggestive of plasmacytoma. After excluding systemic involvement by means of laboratorial and radiological investigations, the patient was referred to radiotherapy department for complete management.

Keywords: Extramedullary, Head and neck, Plasmacytoma, Treatment.

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INTRODUCTION

Plasmacytoma is an extremely rare and discrete solitary mass of neoplastic monoclonal plasma cells, which was first described by Schridde in 1905.[1] Plasmacytoma is the accepted nomenclature for a solitary tumor of plasma cells, located either in bone marrow (medullary plasmacytoma), the solitary plasmacytoma of bone, or outside of bone, and the extramedullary plasmacytoma (EMP).[2] Extramedullary plasmacytoma has been seldom reported and accounts for 4% of all nonepithelial tumors of the upper respiratory tract.[3] While occasionally localized to the gastrointestinal tract, lungs, testes, and skin, it has been previously reported that 80% of EMPs are localized in the head and neck region.[4,5] Extramedullary plasmacytoma is defined as the neoplastic proliferation of plasma cells in soft tissues. It accounts for 3% of all plasma cell tumors. Approximately 90% of EMPs affect the head and neck region, commonly affecting the nasal cavity, paranasal sinuses, tonsillar fossa, and oral cavity. The etiology of this disease is unknown, but chronic stimulation, overdose irradiation, viruses, and gene interaction especially in the reticuloendothelial system have been suggested as etiological factors.[6] The evaluation of a patient with a suspected EMP should include a biopsy of the suspected lesion for tissue histological confirmation, a unilateral bone marrow aspirate and biopsy, and laboratory studies. Imaging should include a metastatic bone survey and either a positron emission tomography/computed tomography (PET/CT) scan or magnetic resonance imaging (MRI) of the entire spine and pelvis.[7-9] The treatment of choice for EMP is surgery and radiation therapy with dose of 40 to 50 Gy over a 4-week period, and the disease is highly radiosensitive. Small lesions may be cured with surgery alone, and no adjuvant radiotherapy (RT) is indicated unless a residual local disease is suspicion.[10]

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CASE SUMMARY

A 55-year-old man from Mandi, Himachal Pradesh,was presented to the Department of ENT at Indira Gandhi Medical College, Shimla, India, on January 15, 2015, with complaints of changes in voice and difficulty in swallowing for last 15 days, which was insidious in onset and gradually progressive in nature; difficulty in swallowing was associated with for both liquid and solid meals both. There was no history of voice abuse and recurrent sore throat. There was no history of fever. There was history of difficulty in breathing intermittently for the last 5 days, which was gradually progressive in nature.

On examination, the patient was afebrile. Throat examination showed congestion with a bulge over the posterior pharyngeal wall which was nontender, about 4 × 5 cm in size, extending toward nasopharynx superiorly and also extending toward left lateral pharyngeal wall. Indirect laryngoscopic examination and posterior rhinoscopy examination could not be possible because swelling was obliterating the view. There was no palpable cervical lympadenopathy. Both nose examination and ear examination were within normal limit.

The patient was admitted into ENT department and conservative treatment started; blood investigation was done which were in normal limit. The patient was posted

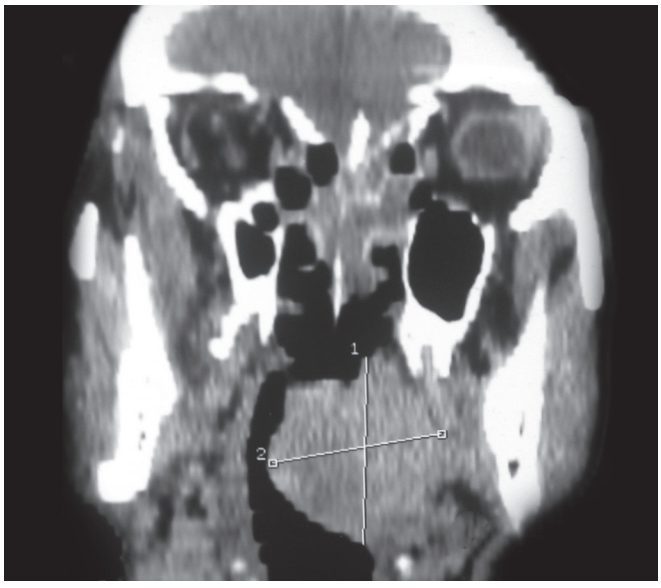

Fig. 1: Computed tomography image showing mass

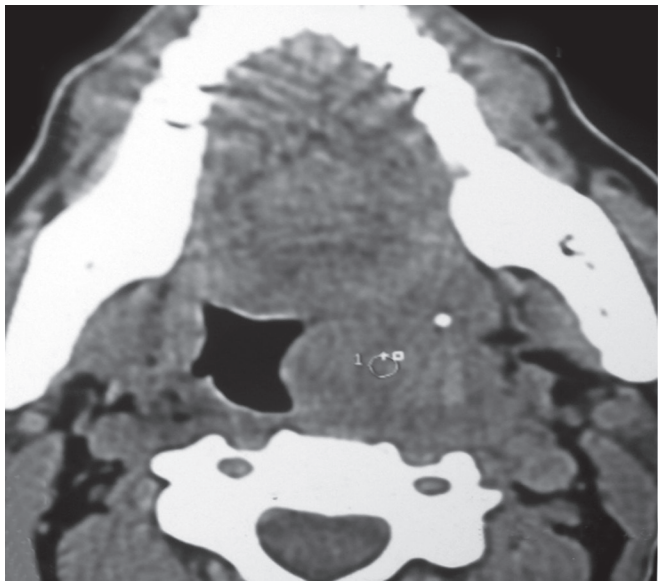

Fig. 2: Computed tomography image showing compromised airway

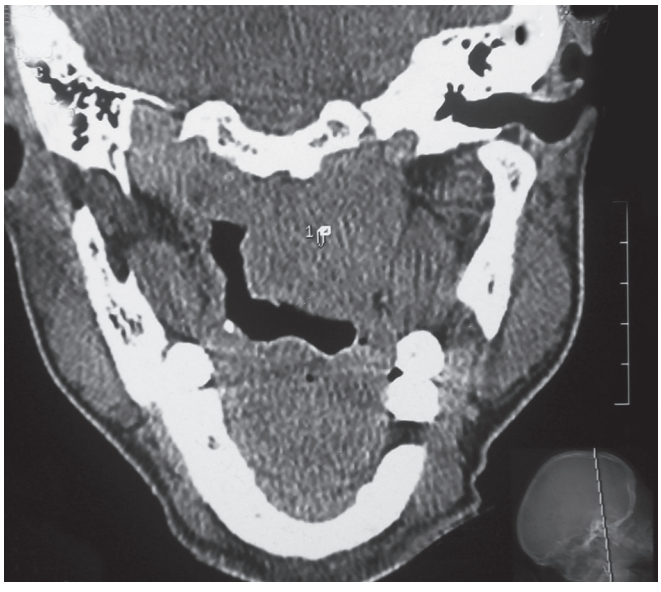

Fig. 3: Computed tomography showing compromised airway

for contrast CT scan of neck and elective tracheostomy for anticipated dyspnea. Ryle's tube was inserted for feeding purpose. Very next day, tracheostomy was done. Contrast CT scan showed a soft tissue density involving the retropharyngeal mass involving parapharyngeal space (Fig. 1) and extending toward nasopharynx and also involving prevertebral space. On CT scan, mass was also compromising the airway significantly (Figs 2 and 3). The patient was posted for excision of this retropharyngeal mass under GA. By giving incision on this mass, mucosal flap were retracted laterally and mass excised, after securing hemostasis incision closed with vicryl sutures.

Peroperatively, there was about 4×4×1 cm, friable, grayish brown mass was excised in toto and sent for histopathological examination (HPE). It was extending superiorly toward nasopharynx and also attached to lateral pharyngeal wall to left side. At postoperative day 10, incision was healthy and Ryle's tube removed and oral feeding started followed by strapping of tracheostomy. The patient was discharged next day and advised to come on follow-up with HPE report.

On follow-up, the patient was in good condition with healthy incision site. Surprisingly, HPE report (Fig. 4) came with rare diagnosis of neck region. It was a round cell tumor with extensive amyloid deposition, suggestive of plasmacytoma (Fig. 5). For this, after excluding systemic involvement by means of laboratorical and radiological investigations, he was referred for RT.

DISCUSSION

Extramedullary plasmacytoma originates from plasma cells with a single class of heavy and light chains in a monoclonal proliferation of B cells. The commonest immunoglobulin expressed by the tumor cells is IgG with kappa chain restriction. Eighty percent of EMPs arise in the soft tissues of the head and neck region. The nasal cavity, paranasal sinuses, and nasopharynx are the most common sites.[11,12] Extramedullary plasmacytoma represent 3% of plasma cell neoplasms. Typical EMP of the upper aero digestive tract is a solitary, sub mucosal, reddish, and sessile or polypoid tumor that is rarely ulcerated. Pedunculated lesions are more often seen in the larynx and pharynx, while sessile tumors have a sinonasal predilection. Friability and softness may indicate a more aggressive lesion.[13]

Tissue biopsy, serum electrophoresis, and radiological skeletal survey with bone marrow study are necessary for diagnosis. The treatment of EMP is surgical resection and RT.[14] In pathology, plasma cells are present with a

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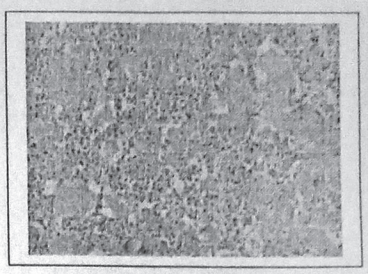
SURGICAL PATHOLOGY REPORT



SPECIMEN	:	Oropharynxal growth biopsy.
CLINICAL HISTORY	:	Hoarseness of voice x 15-20 days. ? Neurofibroma ? Schwannoma.
GROSS	:	Received multiple grey brown to light brown soft tissue bits and pieces together measuring 4.5 x 4 x 1 cm. Cut surface is grey brown to light brown.
MICROSCOPY	:	1. Sections show sheets of plasma cells with abundant amyloid surrounded by foreign body giant cell reaction present separately and focally infiltrating the wall. 2. No evidence of neurofibroma / Schwannoma seen in sections examined.
IMPRESSION	:	**Oropharynxal growth biopsy: Features are suggestive of round cell tumor with extensive amyloid deposition. Suggestive of plasmacytoma.**
ADVISED	:	I. Clinical work up to look for other sites of involvement. II. IHC - CD38, CD138, CD45, CD3, CD20, Kappa and lambda light chains for confirmation.
HISTOPATH NO	:	[24428-24431 : Representative sections]

Dr. Mamta Arora
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Consultant Pathologist

Dr. Abhishek Kumar
MD (PATH)
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Dr. Aparna Gupta
MD(PATH)
Consultant Pathologist

Dr. Shivani Rewri
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Consultant Pathologist

Dr.Tripti Kaur

Dr. Kavita Jain

Dr. Mani Makhija

Dr. Gaurav Sharma

Note: *1. Slides Blocks can be issued only on advise of the referring consultant after a minimum of 48 hours.*
2. Gross specimens will be retained only for a period of 1 month after the date of reporting.
3. Contact histopathology department for any clarification.

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Fig. 4: Detailed HPE report

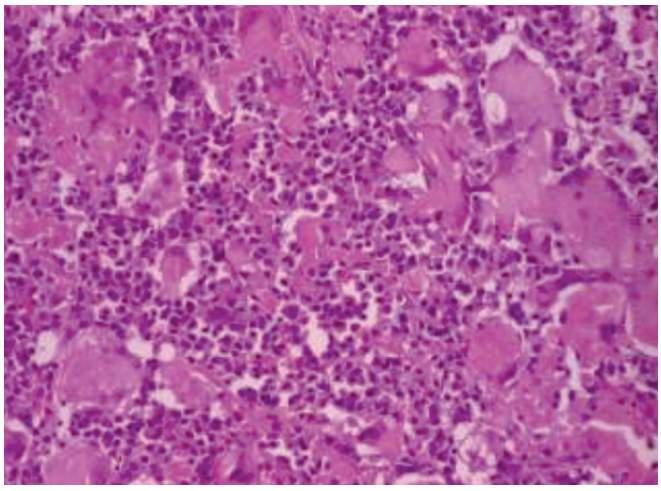

Fig. 5: Slides plasmacytoma

morphologic spectrum ranging from mature forms with abundant cytoplasm and perinuclear halo to highly atypical cells with large nuclei, hyperchromatic clumped chromatin, and prominent nucleoli with scant cytoplasm. Amyloid deposition may be seen in 15 to 38% of EMP. Immunohistochemistry is used to establish clonality.[7,15] Wiltshaw classified soft-tissue plasmacytoma into three clinical stages as follows: Stage I – limited to an extramedullary site, stage II – involvement of regional lymph nodes, and stage III – multiple metastasis.[14] In general, EMPs are considered radiosensitive, with a local control rate of 90 to 100%.[16] A radiation dose of 40 to 50 Gy delivered to the primary site of the EMP in the nasopharynx is usually recommended.[17] Irradiation to the neck is required only in cases with clinically positive cervical node metastasis. In a recent study by Sasaki et al,[18] it was found that RT was quite effective and safe for patients with EMP in the head and neck region. Moreover, RT combined with surgery produced an improved outcome, as determined by survival rates. Although the role of chemotherapy in EMP treatment has not been established, chemotherapy is usually considered for EMPs with high risk factors for local treatment failure (tumor size of > 5 cm) and in cases of refractory disease.[17] Follow-up radiological and electrophoresis assessment is required following treatment to detect recurrence and progression to multiple myeloma, which occurs in 10 to 30% of cases.[19] The 10-year-survival rate is 70%.[17]

CONCLUSION

A literature search revealed no publications supporting the use of surgery alone to treat EMP. In the current case, although the tumor was well defined and thus completely excised, and the patient recovered from the surgery smoothly, subsequent irradiation was recommended. A lifelong evaluation for disease progression is necessary with all plasmacytomas. This includes frequent bone marrow aspirations, skeletal survey with CT or MRI modalities, and serum and urine protein electrophoresis to detect the presence of M proteins.

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