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Solitary Extramedullary Plasmacytoma Masquerading as Osteomyelitis in an Elderly Male - A Rare Case Report

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ABSTRACT

Extramedullary plasmacytoma is characterised by plasma cell proliferation in organs or tissues outside the bone marrow. It originates from B lymphocytes with 60% cases arising in nasal cavity, paranasal sinus and nasopharynx. Extramedullary plasmacytoma corresponds to less than 10% of all plasmacytic tumors representing less than 1% of all head and neck tumors. Typically affects 5th to 7th decade of life having male predominance in Caucasian population. Global incidence is 1 case per 5,00,000 people. This is a rare case of extramedullary plasmacytoma of the soft tissue around the knee a highly uncommon site in an elderly male.

Keywords: Extramedullary plasmacytoma, soft tissue knee

INTRODUCTION

Extramedullary plasmacytoma (EMP) is a disease of clonal plasma cells involving any organ without marrow plasmacytosis. It can arise denovo or during the course of multiple myeloma (1). EMP originates from B lymphocytes with 60% cases arising in nasal cavity, paranasal sinus and nasopharynx. Larynx, oropharynx, skull base, gastrointestinal tract and pelvic cavity are other less common sites(2).

EMP has been classified as true extramedullary lesions that are completely in soft tissue or Paraskel et al with evidence of soft tissue tumour masses developing from underlying bony lesions. They can present as a solitary lesion that is not associated with multiple myeloma or it can be a progressive disease with high risk cytogenetics and resistance to therapy (3). We herein present a rare case of EMP of the soft tissue around the knee in an elderly male.

CASE REPORT

An 84year old elderly man presented with complaints of pain, swelling and discharge over the left knee since four months following history of fall. Patient had a past history of total knee replacement 6 years ago. Ultrasound of left knee suggested synovial hypertrophy with complex knee joint effusion. X ray of left knee showed a failed implant (Figure 1). Operative findings

revealed multiple pus-filled pockets around the implant. Clinically a diagnosis of infective etiology was considered.

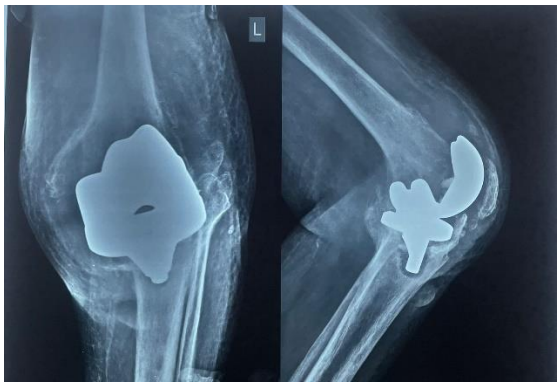

Figure 1) X Ray of the left knee showing failed implant

On gross examination a skin covered soft tissue mass was received measuring 2.7x2.6x1.2cm. Cut surface showed multiple grey brown hemorrhagic areas. (Figure 2)

Figure 2) Cut surface of the soft tissue mass showing grey brown haemorrhagic areas

Microscopically sections studied exhibited epidermis with underlying tumor tissue composed of sheets of round to polygonal abnormal plasma cells with moderate amount of cytoplasm, eccentric nuclei having mild atypia and opened up chromatin. Occasional binucleate plasma cells seen. A diagnosis of solitary extramedullary plasmacytoma was considered. (Figure 3)

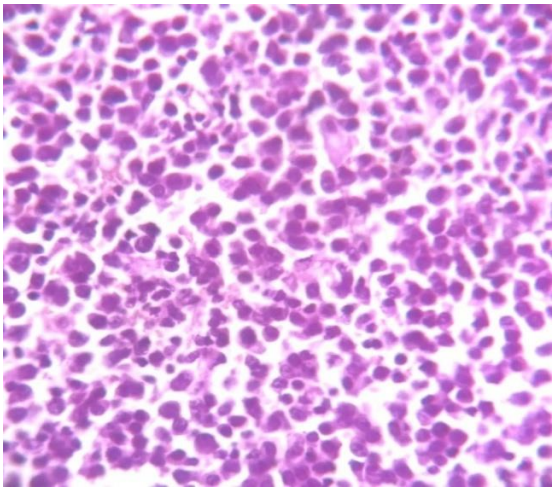

Figure 3) Microscopy showing abnormal proliferation of plasma cells (H & E,400x)

On further work up of the patient complete blood count revealed hemoglobin of 9.5g/dl, total count 23,790 with 90% neutrophils, ESR 120.Peripheral smear showed no presence of plasma cells. Serum calcium and renal function tests were normal. No lytic lesions identified in the long bones.
Immunohistochemistry showed CD138, MUM1, CD56, Lambda positive in tumor cells. Kappa, CD20, CD19 negative in tumor cells confirming the diagnosis of plasma cell neoplasm.

DISCUSSION

Plasmacytic tumors are a group of lymphoproliferative disorders with monoclonal expansion of plasma cells that produce single immunoglobulin molecule. EMP corresponds to less than 10% of all plasmacytic tumors representing less than 1% of all head and neck tumors. It presents as multiple lesions in 10-20% of cases with tumor involving pleura, mediastinum, spermatic cord, ovary, intestine, kidney, pancreas, lung and skin. Typically affects 5th to 7th decade of life having male predominance in Caucasian population. Global incidence is 1 case per 5,00,000 people (4)
Ashish et al reported a rare case of sinonasal extramedullary plasmacytoma with orbital involvement. Etiology of this disease is still unknown but viruses, overdose irradiation, and gene disorders in reticuloendothelial

system have been suggested. The first case of EMP was described in 1905 by Schridde (5)

Accepted criteria for EMP include solitary extramedullary mass of clonal plasma cells, bone marrow plasma cell infiltration (<5%), absence of lytic lesions and absence/low level of serum/urinary monoclonal immunoglobulin (6) Philip et al reported a case of bilateral extramedullary adrenal plasmacytoma (7).We report this case due to rare occurrence of this lesion over the soft tissue around the knee. Hardly any cases reported in the literature.

Rawat et al reported EMP on one side of nasal cavity. Singh et al and Narges et al presented a case report of EMP in the maxillary area Harwood et al reported high rate of conversion to multiple myeloma if EMP involved the bone. Barros et al presented a case report in oral cavity in a 70 year old male (6)

Luis et al reported a case of extraosseous plasmacytoma of parotid gland. Extraosseous plasmacytoma does not show additional lesions on skeletal x ray, plasmacytosis in bone marrow, hypercalcemia, anemia or renal failure. Following treatment approximately 70% of patients remain in complete remission for 10 years. In 25% cases regional recurrence develop and metastasis occurs occasionally (8)

According to case report by Roxanne et al multiple myeloma patient presented with EMP in the maxillary gingiva. Through hematogenous spread EMP can develop in soft tissue in any location of the body in multiple myeloma. The presence of multiple EMP in skin and oral cavity represented relapse and overall poor prognosis of the patient. Treatment modalities for such cases include allogenic stem cell transplant or chemotherapy with lymphoma like regimen(3)

Glasbey et al conducted review among various gastrointestinal manifestations in EMP. Intraabdominal EMP presents as abdominal pain, intestinal obstruction or hydronephrosis. Intraluminal disease shows microcytic anemia, melaena, hematochezia, localised perforation or tenesmus (9)

Jong et al published a case report in which the patient developed soft tissue mass EMP in the back subsequent to radiation therapy for a solitary bone plasmacytoma in the left sinonasal site. Long term follow up is crucial in monitoring for disease recurrence or progression(10)

CONCLUSION

EMP are rare aggressive tumours mainly affecting nasal cavity and paranasal sinuses. Its prognosis is more favourable than multiple myeloma. This case is rare as it highlights a uncommon location of EMP. Long term follow up of the patient is very important due to the risk of relapses reported in the literature.

Declaration by Authors

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Conflict of Interest: The authors declare no conflict of interest.

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