SOLITARY PLASMACYTOMA OF THE CLAVICLE: A CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

Primary tumours of the clavicle are a rarity and overall account for only 0.45% of primary bone tumours. Solitary bone plasmacytomas are similarly a rarity and have distinct prognostic significance in that approximately 50% of untreated lesions will progress to multiple myeloma within 5-years of diagnosis. With regards to the optimal management of a solitary plasmacytoma of bone the attending orthopaedic surgeon must be mindful of the precise location of the lesion. While several studies confirm that a solitary plasmacytomas of bone can be successfully treated with radiotherapy alone, the effect of radiotherapy on the surrounding critical structures must be taken into consideration. With regards to the middle third of the clavicle, the intimate relationship of the inferior surface of the clavicle to the underlying cords of the brachial plexus, is an important consideration. Here the attending orthopaedic surgeon may decide that surgical excision is the treatment of choice with/without adjunctive radiotherapy. We report a case of an adult female patient, who presented to our unit with a relatively painless progressively enlarging mass over the middle third of the clavicle that was subsequently diagnosed to be a solitary plasmacytoma. She was taken to the operating room and successfully managed by *en bloc* resection of the lesion, followed by immediate clavicular reconstruction, with adjunctive radiotherapy being reserved for recurrence. We recommend that even in the context of a known radio-sensitive tumour, the middle third of the clavicle be afforded special consideration when it comes to radiotherapy, and that surgical excision be the treatment of choice for tumours at this specific site.

Key words: Solitary plasmacytoma of bone, Middle third of the clavicle

INTRODUCTION

Less than 1% of bone tumours occur in the clavicle (1). The absence of any significant medullary cavity, and its relatively poor blood supply, are postulated reasons for this paucity (2). While plasma cell dyscrasias comprise a heterogenous group of clinical entities with distinct prognostic implications, they share the same fundamental pathogenesis namely the neoplastic proliferation of monoclonal plasma cells. Included in the spectrum of disease is the most benign, namely Monoclonal Gammopathy of Undetermined Significance (MGUS), the plasmacytoma which has intermediate prognostic significance due to approximately 50% of untreated lesions demonstrating progression to systemic disease within 5-years, and the most clinically aggressive systemic form namely multiple myeloma (3). The World Health Organization (WHO) defines a solitary plasmacytoma of bone as a single lytic bony lesion, comprised of neoplastic monoclonal plasma cells, without associated plasmacytosis on random bone marrow sampling. An additional diagnostic requirement is the absence of any evidence of systemic myeloma which includes renal insufficiency, hypercalcemia, anaemia, a positive bone scan, a monoclonal gammopathy on serum protein electrophoresis, and the presence of Benz Jones protein on urine protein electrophoresis. Solitary plasmacytomas of bone have a predilection to occur in bones of the axial skeleton with active marrow. For this reason, the vertebrae, ribs, skull, pelvis, femur, scapula, and albeit rare the clavicle, are more commonly involved than any bone of the appendicular skeleton (4). Regarding solitary plasmacytomas of the clavicle they are relatively rare. One retrospective series, that enrolled 113 subjects with clavicular tumours, noted that 10/113 (8.8%) subjects had a clavicular plasmacytoma (5). In another study that enrolled 29 subjects with solitary

plasmacytomas, of which 16 were solitary plasmacytomas of bone, reported that only 1/29 (3%) subject had a solitary plasmacytoma of the clavicle (6). We report a case of an adult female patient who presented to our unit with an insidious progressively enlarging mass over the right clavicle that was demonstrated to be a solitary clavicular plasmacytoma. Our case report serves to highlight this rare tumour occurring at a rare site, and as such reminds orthopaedic surgeons to include this in the differential diagnosis of an enlarging clavicular mass.

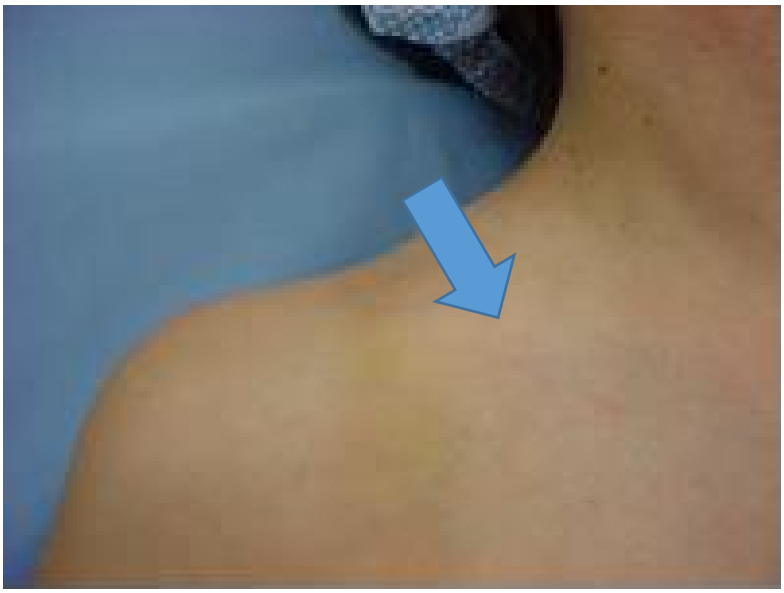
CASE REPORT

A 42-year old female patient presented to our unit with a 6-month history of a progressively enlarging, but relatively painless, right clavicular mass. While she reported the mass to be relatively painless, she did report that she experienced pain when the strap from her brazier pressed on the mass. She was otherwise healthy and had no medical problems. On examination of the mass it was noted to be 4cm by 3cm in size, to have a solid hard consistency, with a discrete edge, and fixed to the middle 1/3rd of the clavicle. There were no overlying skin changes. Neurological examination of her right upper limb was normal (Figure 1).

An X-ray of her right clavicle was performed which showed an intramedullary cortical expanding osteolytic lesion confined to the middle third of the right clavicle. Due to our experience being that most clavicular tumours are malignant, the primary diagnoses that we considered were that the lesion was most likely a chondrosarcoma or an osteosarcoma. On both X-rays the superior cortex was noted to be eroded which we decided to take advantage of to obtain a biopsy (Figure 2).

Figure 1

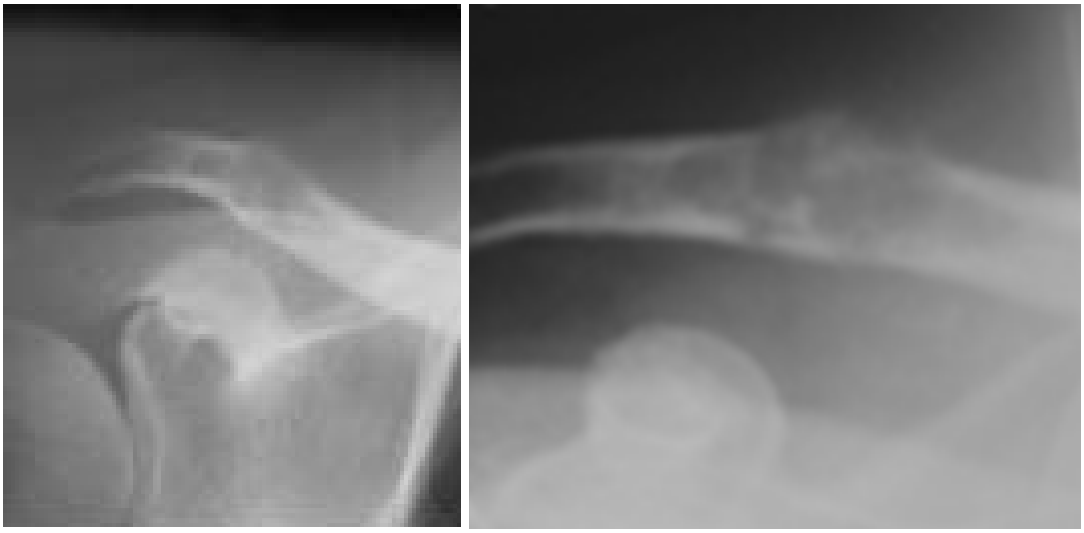
Pre-operative patient photograph showing the right clavicular mass (blue arrow). While the mass can clearly be seen, the obvious absence of overlying skin changes is also appreciated



An MRI of the right clavicle was performed which demonstrated the lesion to be hypointense of MRI T1W imaging, and hyper-intense on MRI T2W imaging. While these were non-specific in terms of establishing the diagnosis, the absence of any obvious infiltration into the surrounding soft tissues was noted and considered favorable.

Figure 2

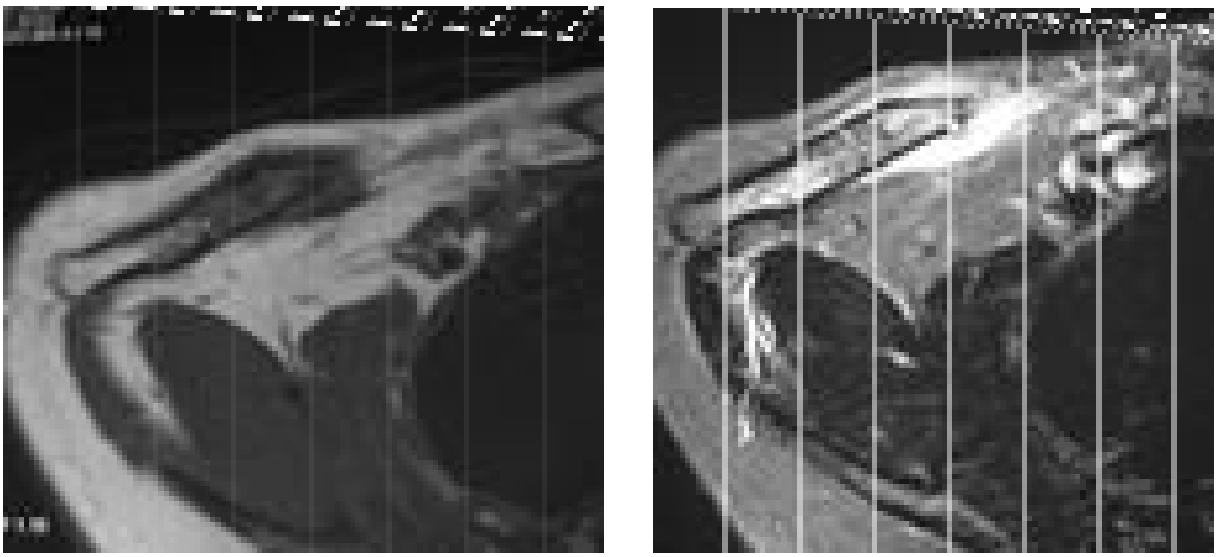
Pre-operative anteroposterior right shoulder X-ray, and 15 degrees superior view X-ray of right clavicle. On these views an intramedullary cortical expanding osteolytic lesion confined to the middle third of the right clavicle is clearly visualized, as is erosion of the superior cortex above the lesion



We performed a core needle biopsy of the lesion which revealed a small round blue cell sheet like infiltrate. On haemoxylin and eosin staining the cells were noted to be oval, with abundant basophilic cytoplasm, and round eccentric nuclei characteristic of mature plasma cells. An absence of intervening parenchyma was further noted. Specific staining performed demonstrated high CD 38 positivity (Figure 4).

Figure 3

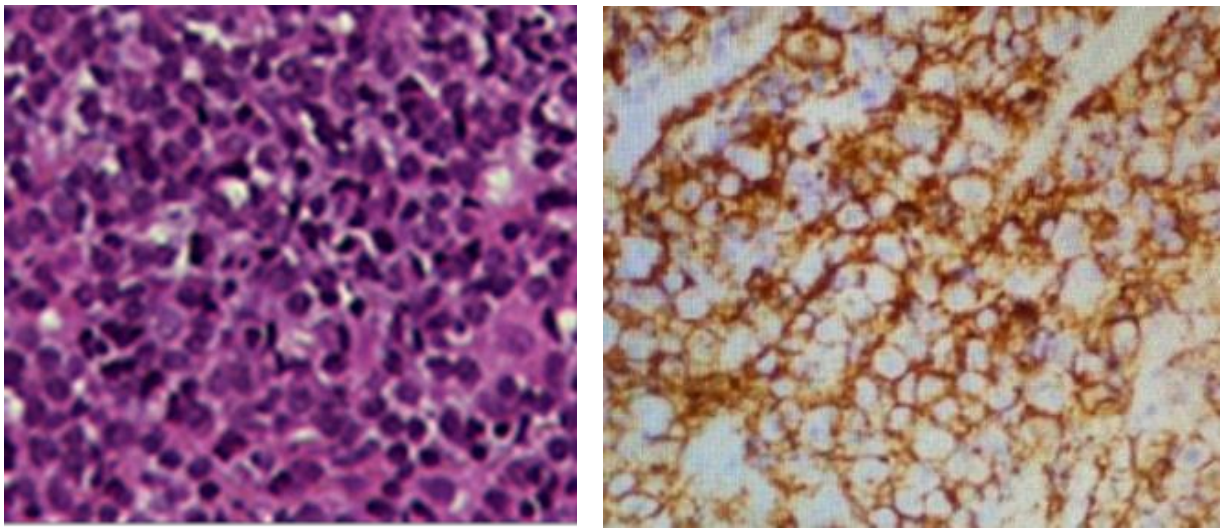
Pre-operative MRI T1W and MRI T2W images of the right clavicle. The lesion is noted to be hypointense on T1W imaging, and hyper-intense on MRI T2W imaging, which is non-specific. The absence of any obvious infiltration into the surrounding soft tissues was considered favorable



We consulted the Haematological Oncology Department to assist in excluding this to be part of underlying more extensive disease. Measurement of serum red blood cell count, platelet count, calcium, urea, and creatine were performed which were all normal. Serum protein electrophoresis was performed which excluded a monoclonal protein peak. Urine protein electrophoresis was performed which was negative for Benz Jones light chain proteins. A bone scan was performed which confirmed no additional lesions to be present on skeletal survey. Random bone marrow biopsies were performed which all confirmed normal bone marrow.

Figure 4

Histological slides of the core needle biopsy, stained on the left with haemoxylin and eosin, showing a small round blue cell sheet like infiltrate, comprised of oval cells with abundant basophilic cytoplasm. Specific staining done, and shown on the right, reveled high positivity for CD 38



We hence diagnosed the lesion to be a solitary plasmacytoma of the clavicle and now needed to decide on the best management. While we realized that solitary bone plasmacytomas can be successfully managed by moderate dose stereotactic radiotherapy comprised of 40-50Gy, administered once daily at 1.8-2Gy per fraction, with reported high local control rates of between 83-96% (7,8), we considered the location of the lesion in the middle third of the clavicle to be unfavorable due to this management incurring a moderate risk of radiation-induced brachial plexopathy. After thorough counselling of the patient, which included discussion of a moderate risk of her later incurring a radiation-induced brachial plexopathy, she declined radiotherapy and opted for surgical excision of the lesion with immediate plating.

Operative procedure: She was taken to the operating room where, post induction of anaesthesia and in a beach chair position, a lazy-S incision was made over the clavicle. Normal clavicle, immediately medial and lateral to the lesion contained within its periosteum, was exposed. After freeing the underlying fascia beneath the lesion, immediately deep to which are the trunks of the brachial plexus, and the fascia under the adjacent clavicle, an osteotome was used to perform an *en bloc* resection of the lesion by cutting through the normal clavicle both medially and laterally. Post *en bloc* removal a further 5mm resection of adjacent normal tissue was performed. The wound was thoroughly irrigated and confirmed to be free of macroscopic disease. Bone graft was harvested from the patient's iliac crest, morselized, and used to fill the bony defect. Double plating of the clavicle, with pre-contoured anatomical locking plates, was then performed (Figure 5). The wound was closed in layers and the skin was closed with skin clips.

An intra-operative fluoroscopic Xray was performed which confirmed restoration of clavicular alignment (Figure 6).

The patient was discharged on the 3rd post-operative day in a broad arm sling. At her 6-week out-patient appointment her wound had healed well, and she reported being satisfied with the outcome. She was discharged for 6-month out-patient review.

Figure 5

Intra-operative patient photograph showing double plating of the patient's clavicle, utilizing pre-contoured anatomical locking plates. The morselized bone graft used to fill the bony defect can be seen (blue arrow)

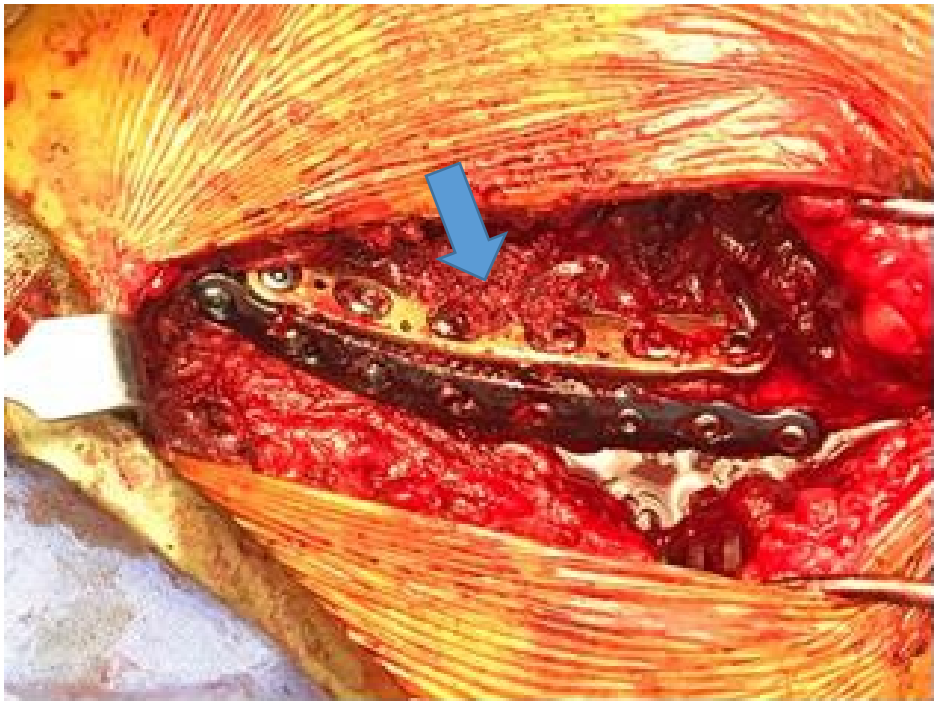
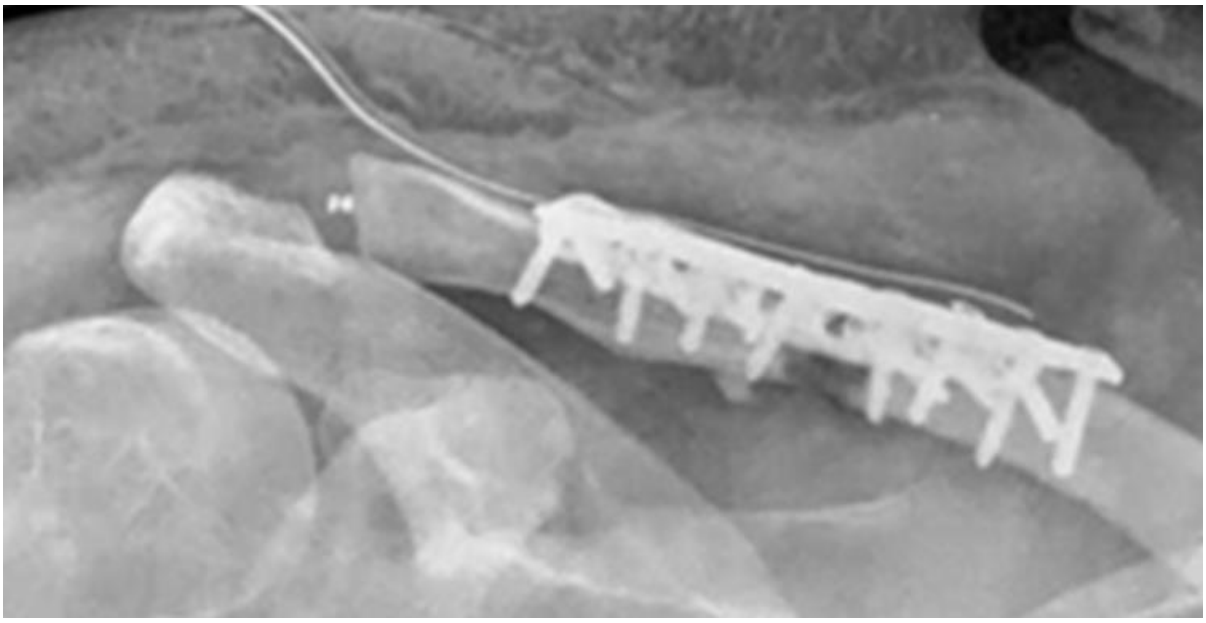


Figure 6

Intra-operative fluoroscopic X-ray confirming restoration of clavicular alignment. The double plating, with pre-contoured anatomical locking plates, can be seen



DISCUSSION

One retrospectively series, that enrolled 1300 patients with primary bone tumours, reported that only 0.45% were clavicular (9). Another study, that enrolled 29 subjects with solitary plasmacytomas, reported that only 1/29 (3%) was clavicular (6). A further study, that enrolled 32 patients with solitary plasmacytomas, similarly reported only 1/32 (3%) to be clavicular (10). These studies hence confirm not only the relative rarity of primary bone tumours of the clavicle, but also the rarity of this to be a solitary bone plasmacytoma. Common tumours of the clavicle, reported in a retrospective series of 113 patients, are eosinophilic granulomas which occurred in 21/113 (18.8%) subjects, metastases which occurred in 17/113 (15%) subjects, Ewing's sarcoma which occurred in 10/113 (8.8%) subjects, osteosarcoma which occurred in 9/113 (8%) subjects, aneurysmal bone cyst which occurred in 10/113 (9%) subjects, and chondrosarcoma which occurred in 5/113 (4.4%) subjects (5).

Several studies note that radiotherapy alone results in successful local control of primary bone plasmacytomas (7,8,11,12). One case report gives an account of a solitary plasmacytoma of the medial end of the clavicle that was successfully treated with surgical excision followed by adjunctive radiotherapy (13). In our patient the solitary bone plasmacytoma was in the middle third of the clavicle, directly deep to which are the cords of the brachial plexus, and hence we decided there to be a moderate risk of her incurring a radiation-induced brachial plexopathy. After thorough counselling of the patient, it was agreed that we would offer her surgical excision alone, and reserve adjunctive radiotherapy for recurrence, thereby completely avoiding this risk. While radiotherapy alone is reported to be successful, another study notes surgical excision to be the treatment of choice for the primary management of solitary bone plasmacytomas, where possible. Here the surgical options include subtotal, or total clavulectomy with or without re-construction, which reportedly results in no to little disability (4-16).

CONCLUSIONS

Our case report gives an account of an adult female patient who presented with, what was later diagnosed to be, a solitary plasmacytoma of the clavicle. After consideration of the precise location of the lesion, in the middle third of the clavicle in proximity to the cords of the brachial plexus, it was decided that we would manage her by *en bloc* resection. While solitary plasmacytomas of the bone are known to respond favorably to radiotherapy alone, we recommend that with regards to the clavicle, consideration need to be made for the actual site of the tumour, with the medial third of the clavicle being distinct in that immediately deep to it likes the cords of the brachial plexus. In these cases, we recommend surgical excision to be the treatment of choice, and that adjunctive radiotherapy be reserved for recurrence.

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REFERENCES

1. Kapoor, S., Tiwari, A. and Kapoor, S. Primary tumours and tumorous lesions of clavicle. *Int Orthop.* 2008; **32**: 829–834.
2. Pressney, I. and Saifuddin, A. Percutaneous image-guided needle biopsy of clavicle lesions: a retrospective study of diagnostic yield with description of safe biopsy routes in 55 cases, *Skeletal Radiol.* 2015; **44**:497–503.
3. Castillo, J. Plasma cell disorders. *Prim Care Clin Office*. 2016; **43**(4):677–691.
4. Jaffe, E.S., Harris, N.L., Stein, H. and Vardiman JW. World Health Organization classification of tumours. Pathology and genetics of tumors of the haematopoietic and lymphoid tissues. Lyon 2001, IARC press.
5. Priemel, M.H., Stiel, N., Zustin, J., Luebke, A.M., Schlickewei, C. and Spiro, A.S. Bone tumours of the clavicle: Histopathological, anatomical, and epidemiological analysis of 113 cases. *J Bone Oncology.* 2019. https://doi.org/10.1016/j.jbo.2019.100229.
6. Basavaiah, S.H., Lobo, F.D., Philipose, C.S., Suresh, P.K., Sreeram, S., Kini, H., Sahu, K.K. and Prasad, K. Clinicopathological spectrum of

solitary Plasmacytoma: a single center experience from coastal India. *BMC Cancer.* 2019;**19**:801. https://doi.org/ 10.1186/s 12885-019-5976-7.
7. Mayr, N.A., Wen, B.C., Hussey, D.H., Burns, C.P., Staples, J.J. and Doornbos, J.F. The role of radiation therapy in the treatment of solitary plasmacytomas. *Radiother Oncol.* 1990; **17**:293-303.
8. Holland, J., Trenkner, D.A., Wasserman, T.H. and Fineberg, B. Plasmacytoma. Treatment results and conversion to myeloma. *Cancer.* 1992; **69**: 1513-17.
9. Klein, M.J. and Siegal, G.P. Osteosarcoma: anatomic and histologic variants. *Am J Clin Pathol.* 2006; **125**(4):555–81.
10. Shih, L.Y., Dunn, P., Leung, W.M., Chen, W.J. and Wang, P.N. Localized plasmacytomas in Taiwan: comparison between extramedullary plasmacytoma and solitary plasmacytoma of bone. *Br J Cancer.* 1995; **71**:128-133.
11. Mill, W.B. and Griffith, R. The role of radiation therapy in the management of plasma cell tumors. *Cancer.* 1980; **45**(4):647-652.
12. Barzenje, D.A., Kolstad, A., Ghanima, W. and Holte, H. Long-term outcome of patients with solitary plasmacytoma treated with radiotherapy: A population-based, single-center study with median follow-up of 13.7 years. *Hematol Oncol.* 2017:1-7. Doi 10.1002/ hon. 2415.
13. Dhyani, A., Shankar, A., Gupta, A., Kumar, R., Chatterjee, K. and Majumdar, A. Solitary plasmacytoma of clavicle: A case report. *IJCRI.* 2013; **4**(8):440–443.
14. Ren, K., Wu, S., Shi, X., Zhao, J. and Liu, X. Primary clavicle tumors and tumorous lesions: a review of 206 cases in East Asia. *Arch Orthop Trauma Surg*. 2012; **132**:883–889.
15. Angelini, A., Mavrogenis, A.F., Rimondi, E., Rossi, G. and Ruggieri, P. Current concepts for the diagnosis and management of eosinophilic granuloma of bone. *J Orthop Traumatol*. 2017; **18**:83–90.
16. Martin, J.R., Mazzini, J.P., Fernandez, R.V., Ciruelos, R.M. and de la Mano, A.C. Ewing sarcoma of clavicle in children: report of 5 cases. *J Pediatr Hematol Oncol*. 2009; **31**:820–824.