CASE REPORT AND REVIEW OF LITERATURE

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Small cell variant of osteosarcoma of the patella: A rare case report with review of literature

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ABSTRACT

Patella is an uncommon site for the occurrence of bone tumor. Benign tumors are more common as compared to malignant ones. Osteosarcoma of the patella is a very rare tumor with only a few cases reported in the literature. Here, we present a rare case of a small-cell variant of osteosarcoma of the patella. A 12-year-old boy presented to us with pain and swelling over his left knee for 6 months. On X-ray, expansion with osteosclerotic and osteolytic lesions was found in the patella and a suspected diagnosis of osteosarcoma was made. The patient was managed by excisional biopsy of the patella and V–Y plasty of the quadriceps tendon. Results of histopathological and immunohistochemistry analyses were consistent with osteosarcoma of small cell variant. The patient was referred to a higher center after the operation for radiotherapy and chemotherapy. On the final follow-up, after 1 year, the patient was found to have expired. Osteosarcoma of the patella is a rare condition. Further studies with a number of cases and longer duration of follow-up are warranted to obtain better insight that can aid in devising management protocol, prediction of survival, and functional outcome of different surgical interventions for these patients.

Key words: Osteosarcoma; Patella; Patellectomy; V–Y plasty of quadriceps; Small cell variant

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INTRODUCTION

Patella is a sesamoid bone. It is a very rare site for any type of bone tumor, with benign tumor being more common than the malignant form. Tumor and tumor-like lesions arising primarily from the patella are extremely rare, accounting for about 0.12% of all primary bone tumors. Because of sesamoid origin and size of the patella, typical features of bone tumors, such as periosteal new bone formation, may be missing. This limits the specificity of imaging in distinguishing between benign, malignant, and tumor-like lesions. The patella can be a variety of benign bone tumors including giant cell tumor (GCT), chondroblastoma, aneurysmal bone cyst, enchondroma, osteoid osteoma, osteochondroma, osteoblastoma, osteomyelitis, and simple bone cyst. Other less common benign lesions are osseous hemangioma, lipoma, osteitis fibrosa cystica, angioleiomyoma, intraosseous gout, and intraosseous ganglionic cyst. The most common benign bone tumor of the patella is giant cell tumor representing about 30% followed by chondroblastoma which accounts for nearly 16% of cases. The rest of the patellar tumors have a relatively very low incidence rate. Whereas malignant tumors of the patella are much less common, patellar metastasis from primary tumors are frequent. Primary malignant neoplasms with bony metastases such as breast, thyroid, lung, prostate, and renal cell cancers are expected to be the most common cause of patellar bone metastasis. A slightly higher incidence might be observed with lung and renal cell carcinoma given their tendency to cause acrometastasis (metastasis distal to knee and elbow). Among the primary malignant bone tumors, which include osteosarcoma, chondrosarcoma,

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hemangioendothelioma, leiomyosarcoma, angiosarcoma, Ewing's sarcoma, malignant fibrous histiocytoma, and lymphoma, osteosarcoma is the most common and is mostly found around the knee – proximal tibia and distal femur at the metaphyses of long bones – but the involvement of the patella is very rare. Osteosarcoma represents 6% of the total of the patellar tumors, whereas chondrosarcoma accounts for 20–27% of the malignant tumors of the patella. The most common presentation of tumor of the patella is anterior knee swelling. Because tumor of the patella is a rare differential diagnosis for anterior knee pain, its diagnosis is much delayed. Nonetheless, despite the patella being a rare site for bone tumorous growth, chronic anterior knee pain or swelling should always arouse suspicion for such a possibility.[1-6]

CASE PRESENTATION

A 12-year-old boy presented at the outpatient department of our hospital with painless swelling, persisting for 6 months. The swelling was gradually increasing in size. The patient had a history of loss of weight and appetite but did not report trauma and had no history of cough, hemoptysis, chest pain, or tuberculosis. There was no history of morning stiffness of any other joint. A few months later, the patient started complaining of anterior knee pain, which was relieved by conservative management; however, the swelling persisted.

On physical examination, diffuse swelling of size about 8 cm × 9 cm originating from the patella was noted over the anterior part of the patella (Figure 1). The area was firm, tender, and palpable with raised local temperature and no joint effusion. There was a slight restriction in knee movement and gross wasting of the left thigh muscle. No distal vascular or neurological deficit was noted. X-ray imaging showed an enlarged patella with osteosclerotic and osteolytic lesions with cortical thinning/breach in the patella (Figure 2). Essential blood investigations were performed. Serum alkaline phosphatase and serum lactate dehydrogenase levels were significantly raised. There was no marked increase in the levels of inflammatory markers. Suspected diagnosis of osteosarcoma was made, and excisional biopsy of the lesion was planned. All contraindications of surgery were ruled out and written consent was taken from the patient's attendant after explaining the procedure.

The patient was managed by excisional biopsy of the patella and extensor mechanism repair. Surgery was performed under spinal anesthesia; an anterior midline incision was given over the left patella, and subcutaneous tissue

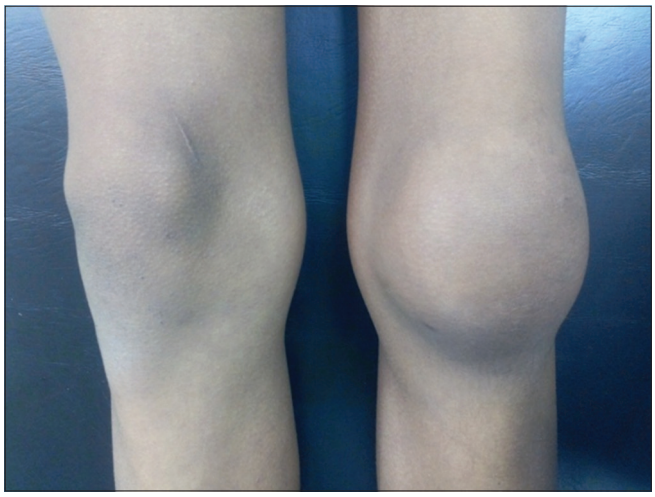

Figure 1: Osteosarcoma of the left patella presenting as an anterior knee swelling

Figure 2: Lateral radiograph of the knee showing enlarged patella with osteosclerotic and osteolytic lesions and cortical thinning/breach

and soft tissue were dissected. The quadriceps tendon from the superior pole, and ligamentum patellae, from the inferior pole of the patella, were detached, and the entire patella was removed as *in toto* mass (Figures 3-5). On gross intraoperative examination, no involvement of ligamentum patellae and quadriceps tendon or synovium of the knee joint was observed. The extensor mechanism repair was performed by suturing the ligamentum patellae and quadriceps tendon together using a V-Y plasty of quadriceps tendon in extension, and a cylinder plaster slab was applied. The excised patella was sent for histopathology and immunochemistry.

On histopathological examination, fibrocartilaginous tissue, skeletal muscle bundles, and bone infiltrated by the tumor dispersed in sheets of small round cells with intervening dense fibrous septa and extracellular eosinophilic matrix (osteoid) were evident. The tumor

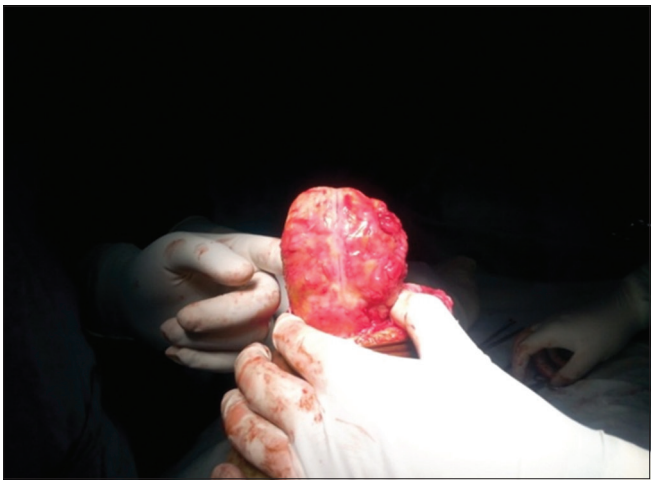

Figure 3: Patella was exposed and separated as *in toto* mass

Figure 4: Excised patella with tumor mass measuring about 8 cm × 9 cm, showing posterior articular surface of it

Figure 5: Intraoperative photograph of left knee after patellectomy

cells were small to medium in size and had sparse eosinophilic cytoplasm and hyperchromatic irregular nuclei. Mitotic and necrotic areas and foci of mature hyaline cartilage were visible in histological sections. The sections were positive for vimentin, pan-CK, and CD-99. These findings were consistent with the rare small-cell variant of osteosarcoma.

Postoperatively, the patient was kept non-weight bearing. On the 14th day, the stitches were removed; and the cylinder plaster of the slab was reapplied. Thereafter, the patient was discharged without any complications and referred to a higher center for chemotherapy and radiotherapy. On follow-up after 1 year, the patient was found to have expired.

DISCUSSION

Among the various causes of anterior knee pain, patellar tumor represents a rare etiology. As such, the diagnosis of this tumor is often much delayed. The most common presentation of a patellar tumor is knee swelling. Chronic anterior knee pain or swelling should always arouse suspicion of a patellar tumor.[1-4] With only 35 cases reported in literature, the incidence of osteosarcoma of the patella is about 6% of all patellar tumors.[7-10] Due to such a low incidence, the literature on the management of patellar tumor is scarce, with only 20 cases having been published as of date (Table 1).

The patella is an unusual site for primary bone tumors and a majority of lesions in the patella are benign. The incidence of malignant tumors (27%) of the patella is less as compared to that of the benign tumors (73%).[1] In the Mayo Clinic series, only one case showing osteosarcoma of the patella among 1649 cases of osteosarcoma was reported.[27] The ossification in the patella is considered similar to the epiphysis of the long bone. Reasonably, GCTs and chondroblastomas, which are regarded as epiphyseal tumors, are the most common tumors in the patella.[1,22,28,29]

Depending on the nature of tumor mass and stage of growth, there are various modes of treatment of osteosarcoma of the patella; these include patellectomy, above-knee amputation, patellectomy with extensor mechanism repair by various grafts or patellectomy without extensor mechanism repair, extra-articular knee resection, and an above-knee amputation. Surgical excision is the treatment of choice for osteosarcoma at any site. Whether to perform an extensor mechanism reconstruction or not is an important question after extensive patellectomy. There are various modes of reconstruction of the extensor mechanism. A few surgeons have attempted extensor mechanism reconstruction using different techniques. Cho et al., used an allograft patella, Aoki et al., performed

Table 1: Details of reported cases of osteosarcoma of the patella

Case	Author	Year	Age/Sex	Presenting interval	Initial treatment	Final treatment	Remarks
1	Neumann[11]	1871	NA	NA	NA	Tumor excision	Periosteal osteosarcoma
2	Hayem and Graux[12]	1874	22/ Female	NA	NA	NA	Osteosarcoma
3	Wild[13]	1882	30/ Female	NA	NA	Above knee amputation	Osteosarcoma
4	Parker[14]	1896	13.5/ Female	NA	NA	Local resection	Periosteal osteosarcoma
5	Creite[15]	1906	42/Male	NA	NA	Local removal and excision of knee joint	Osteosarcoma with giant cells
6	Schmidt[16]	1907	45/ Female	NA	NA	Above knee amputation	Small cell osteosarcoma
7	Bianchetti[17]	1926	NA	NA	NA	Local excision	Small cell osteosarcoma
8	Bellini[18]	1934	23/Male	NA	NA	Above knee amputation	Primary endothelial osteosarcoma
9	Goodwin[19]	1961	24/Male	6 months	Patellectomy	Above knee amputation	Primary osteosarcoma
10	Nagai et al.[20]	1993	34/ Female	7 months	Steroid injection	Patellectomy	Chondroblastic osteosarcoma
11	Okada et al.[21]	1994	54/Male	3 months	Chemotherapy	Patellectomy by Miyakawa method	Post-radiation osteosarcoma
12	Ferguson et al.[22]	1997	18/ Female	Not available	Patellectomy	Knee resection+prosthesis	Associated with Rothmund Thomson syndrome
13	Ishikawa et al.[23]	2000	35/ Female	Not available	Not available	Not available	Associated with Werner syndrome
14	McGrath et al.[24]	2006	53/ Female	3 months	Steroid injection	Chemotherapy+knee resection	Osteogenic sarcoma of patella spread to lateral meniscus after arthroscopy
15	Shehadeh et al.[10]	2008	22/Male	Not available	Physical therapy	Amputation	Telangiectatic osteosarcoma
15	Cho et al.[7]	2009	53/ Female	3 years	Not available	Reconstruction after patellectomy	Biologic reconstruction with allograft
16	Singh et al.[4]	2009	18/Male	3 years	Not available	Patellectomy	Presented case series of tumor and tumor-like lesion of the patella
17	Chida et al.[8]	2012	31/ Female	2 years	Preoperative chemotherapy	Above knee amputation+chemotherapy	Fibroblastic osteosarcoma
18	Aoki et al.[9]	2014	47/Male	1 year	Not available	Chemotherapy+Patellectomy	Conventional osteosarcoma
19	Agarwal et al.[25]	2020	25/Male	6 months	NSAID	Chemotherapy+patellectomy without extensor mechanism repair	Osteoblastic osteosarcoma
20	Kuchhal and Lamture[26]	2023	54/ Female	10 years	Not available	Not available	Chondroblastic type osteosarcoma
21	The current case	2024	12/Male	6 months	NSAID	Reconstruction after patellectomy	Small cell variant of osteosarcoma

a free anterolateral thigh flap, and Ferguson et al., used fascial slings and rotational gastrocnemius flap for extensor mechanism reconstruction.[7,9,30] However, all these procedures may have an unpredictable outcome and are fraught with several problems, such as the possibility of rejection of an allograft, infection due to the use of replacement prostheses, or necrosis of the tissue used for reconstruction due to chemotherapy. There is a risk of a delay in the post-operative chemotherapy in the event of any wound-related complication from a more complex surgery, which severely affects survival. Out of the twenty cases reported in the literature, six patients underwent above-knee amputation, two had knee resection, two underwent patellectomy with extensor mechanism reconstruction, and one patient was managed by patellectomy without extensor mechanism repair. In

the current case, the patient was managed by patellectomy and extensor mechanism repair using V-Y plasty of the quadriceps tendon.

The delay in diagnosis of a patellar tumor is associated with a high morbidity rate. A complete eradication of the disease should be the aim to improve survival even at the cost of functional rehabilitation and not vice versa. No major knee dysfunction results after total patellectomy have been reported earlier.[31,32]

Limb salvage, particularly reconstructing extensors of the knee, has been debated in the literature. As it is a rare site of osteosarcoma, only a few cases have been managed by limb salvage and reconstruction of the extensor mechanism. Agarwal et al., reported the management of a diagnosed case of osteosarcoma of the patella by excision of the patella and some part of the quadriceps tendon and ligamentum patellae, they managed the gap was created, thereby, without extensor mechanism repair and cylinder slab for 6 weeks with good results, without any extensor lag, at 1-year follow-up.[25] Okada et al., manage one case of post-radiation osteosarcoma of the patella by Miyakawa's method (Miyakawa's patellectomy realigns the extensor mechanism, with the proper tension, and centers the functional pull of the quadriceps tendon and patellar ligament; a superficial strip of the quadriceps tendon is pulled distally to fill the void that was left by the removal of the patella and to maintain proper length and the musculotendinous portion of the vastus lateralis and vastus medialis are advanced over this defect in the midline and are sutured to the quadriceps tendon).[21,33]

We have, herein, presented the uncommon case of patellar osteosarcoma in a young male, which was managed by wide excision with extensor mechanism reconstruction by V-Y plasty of the quadriceps tendon. Because the patient expired after 1 year of surgery, we refrain from commenting on the functional outcome of the management strategy employed by un-considering the short period of survival after tumor resection.

CONCLUSION

In patellar osteosarcoma, chemotherapy together with optimal excision for obtaining tumor-free margins, comprising complete patellectomy followed by extensor mechanism reconstruction using V-Y plasty of the quadriceps tendon is a viable option. Further studies with more number of cases and longer follow-up duration are needed for a better understanding of this rare condition, for devising management strategies and assessing their functional outcomes, and for the prediction of patient survival.

CONSENT

The authors certify that they have obtained all appropriate patient consent forms. In the form, the parents have given consent for his images and other clinical information to be reported in the journal. The patient understands that their name and initials will not be published and due efforts will be made to conceal their identity.

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