

Intraosseous schwannoma of the proximal humerus with pathologic fracture

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Case report

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Abstract

Purpose: Intraosseous schwannomas are extremely rare in the humerus, and less than 5 cases have been reported previously in the literature. This is the first report of its origin in the proximal humerus with pathologic fracture. We herein present this case for discussing the reason for its rarity and sharing our experience of management.

Case presentation: A 55-year-old female patient who presented with pain in the right shoulder, which caused by tripping and falling over a board. Radiographs, computed tomography (CT) and magnetic resonance imaging (MRI) showed considerable tumor in proximal humerus, which connected with a fracture. For this suspected tumor, we performed two operations. Pathological examination demonstrated typical picture of a schwannoma, showing whorls and interlacing fascicles of schwannoma spindle cells. Immunohistochemistry, The tumor cells were diffusely positive for S-100 protein, SOX-10 and CD68, while they were completely negative for desmin, DOG-1, AE1/AE3 and P63. The ki-67 index was about 10%. No mitoses or features of malignancy were identified. Finally, a diagnosis of benign schwannoma with focal of actively proliferated cells was made.

Methods: The treatment for intraosseous neurilemmomas with pathologic fracture include excisional biopsy, curettage, bone allograft, and fracture fixation.

Results: The patient recovered well. After the surgery, the patient gradually regained mobility and pain subsided. There was no recurrence after 6 months follow-up by X-ray.

Conclusion: In our case, the tumor with higher CD68 staining were likely to demonstrate that the tumor volume increase is not only based on cell proliferation, but also intratumoral hemorrhage, vascularization, and inflammation, which may be produce rarefaction of the bone and lead to bone fracture after a trivial trauma.

Key Points

1. Intraosseous schwannomas are extremely rare in the humerus, and less than 5 cases have been reported previously in the literature.
2. This is the first report of its origin in the proximal humerus with pathologic fracture. Owing to its scarcity, with the consent of the patient, we herein present this case for discussing the reason for its rarity and sharing our experience of management.

Importance Of The Study

In this study, we performed several immunohistochemical stainings, including S-100 protein, SOX-10, CD68, desmin, DOG-1, AE1/AE3, P63 and Ki-67, which are used as ancillary tests for diagnosis of the tumor. It is remarkable that the tumor in our case with higher CD68 staining. Generally, the degree of inflammation measured by the expression of CD68 showed a positive significant correlation with tumor

size and tumor growth index. This is inflammatory in nature and may contribute to the pathogenesis of schwannoma. In our case, the tumor with higher CD68 staining were likely to demonstrate that the tumor volume increase is not only based on cell proliferation, but also intratumoral hemorrhage, vascularization, and inflammation, which may be produce rarefaction of the bone and lead to bone fracture after a trivial trauma.

Introduction

Schwannoma is a neurogenic tumor that arises from a local proliferation of schwannoma cells in the peripheral, cranial, or visceral nerve. Intraosseous schwannomas account for less than 0.2% of primary bone tumors, and the most common site for intraosseous schwannoma is the mandible. To our knowledge, Intraosseous schwannomas are extremely rare in the humerus, and less than 5 cases have been reported previously in the literature. This is the first report of its origin in the proximal humerus with pathologic fracture.

Case Presentation

A 55yearold female patient was referred to our hospital with right shoulder pain caused by tripping and falling over a board on the same day. She had pain and restriction of movements of her right shoulder joint. Her history was notable for having antecedent pain in the right shoulder with restriction of overhead movements for about 1 year. She denied previous shoulder injury, or surgery. On presentation, The patient complained about right shoulder pain, swelling and ecchymosis. On palpation, tenderness was noted all around the shoulder. The skin of the shoulder was intact, and there was no evidence of warmth, erythema, or induration. Both neurological and vascular examinations were normal.

Radiographs of the right shoulder showed a large, well-defined osteolytic tumor, which gives an appearance of endosteal scalloping and trabeculated contours on the edge of the bone lesion, involving the proximal humerus(Fig. 1A, Fig. 1.B). No significant periosteal reaction was present, and no soft tissue mass or central calcifications were found.

Computed tomography (CT) showed considerable welldefined osteolytic lesion with cortical ballooning and thinning. The density of the tumor was homogeneous and had invaded into the cortex. A intramedullary mass elevating the periosteum was seen at the axial slice bone window and a fracture was seen at the coronal slice bone window (Fig. 1C, Fig. 1.D).

Magnetic resonance imaging (MRI) of the right shoulder showed the tumor had invaded into the cortex and the bone lesion with associated soft tissue edema. The lesion was isointense to muscle on T1-weighted images (Fig. 2A, Fig. 2C), and hyperintense on T2-weighted images (Fig. 2B, Fig. 2D). It was noted involving anterior, medial, posterior aspect of proximal humerus. The shoulder joint space was uninvolved.

Because of uncertainty regarding the histological origin of the tumor, we performed an open biopsy. The operation was performed using a standard deltopectoral approach to access the proximal humerus. Intraoperative the anterolateral cortex of the the proximal humerus was elevated (cortical flap). After exposing and removing the upper bony sheath, we found that the tumor was a well-encapsulated soft grayish-yellow colored mass (Fig. 3A). The pathological tissue was removed and sent for histopathological analysis.

Pathological examination demonstrated typical picture of a schwannoma, showing whorls and interlacing fascicles of schwannoma spindle cells. (Fig. 4A,B). Immunohistochemistry for S-100 protein, SOX-10, CD68, desmin, DOG (discovered on gastrointestinal stromal tumors)-1, AE1/AE3, P63, Ki-67 was performed. The tumor cells were diffusely positive for S-100 protein, SOX-10 and CD68(Fig. 5A,B,C), while they were completely negative for desmin, DOG-1, AE1/AE3 and P63 (Fig. 6A,B,C,D). The ki-67 index was about 10%(Fig. 5D). No mitoses or features of malignancy were identified. Finally, a diagnosis of benign schwannoma with focal of actively proliferated cells was made.

Then the patient was scheduled 10 days after the biopsy for an extended curettage, bone allograft, and fracture fixation, using the same approach. Intraoperative, a complete and meticulous curettage was performed, the cavity was filled with allograft bone(Fig. 3B), and the pathologic fracture was fixated with a proximal humerus locking plate(Fig. 3C,D).

The patient recovered well. After the surgery, physiotherapy was started the day after the operation and a sling was given for comfort. The patient gradually regained mobility and pain subsided. There was no recurrence after 6 months follow-up by X-ray.

Discussion

Schwannoma is a benign tumour developing from Schwann cells, first described by Verocay in 1908, and shows a predilection to the myelinated nerves, especially the sensory nerves[1]. Intraosseous schwannomas are rare benign neoplasms of the bone of which fewer than 200 cases have been described in the world literature[2]. In all of these, the majority of intraosseous schwannomas have been located in the mandible. Kito M and his colleagues have suggested that the high frequency in the mandible is not because of the long intraosseous course, but because the mandibular nerve consists of sensory nerves of the trigeminal nerve origin[3]. On the contrary, most intraosseous nerves are non-myelinated and participate in vasomotor functions. This may be the cause of Schwannoma rarely arising in the bones of the extremities[4], such as the humerus. Up until now, less than 5 cases[5, 6, 7, 8] have previously been reported in the humerus. In this paper, we present the first case report of an intraosseous neurilemmoma affecting the proximal humerus with pathologic fracture.

Clinically, Intraosseous schwannoma may be present for years before becoming symptomatic, and pain may be present in about 50% of the cases, whereas no symptoms are present in about 25%[9]. They are often discovered as an incidental finding[10]. In our case, the tumor was found after the incidence of a

trivial trauma, even though the patient having antecedent pain in the right shoulder with restriction of overhead movements for about 1 year.

Preoperatively, the possibility of intraosseous schwannoma was not considered in our case because of its rarity and nonspecific clinical and radiological findings. The typical radiographic appearance of intraosseous Schwannoma is a well-defined, cyst-like lesion with a thin sclerotic border[11], Computed tomography facilitates the detection and delineation of these tumors, and MR imaging shows an isointense signal to muscle on T1-weighted images and homogeneously or heterogeneously hyperintense to fat on T2-weighted images[12]. These characteristics were all manifest in our case. However, these findings are non-specific[13]. The final diagnosis was not made until histologic examination of tissue obtained.

Pathologically, intraparenchymal schwannomas have similar appearances to soft tissue schwannoma and demonstrate two types of cell arrangements, alternating cellular (Antoni A) and myxoid (Antoni B) areas[14]. Immunohistochemical staining can facilitate the diagnosis. In this study, we performed several immunohistochemical stainings, including S-100 protein, SOX-10, CD68, desmin, DOG-1, AE1/AE3, P63 and Ki-67, which are used as ancillary tests for diagnosis of the tumor. Diffuse immune reactivity for S100 protein and SOX-10 is indicative of schwann cell origin. Negative staining for smooth muscle marker desmin[15], gastrointestinal stromal marker DOG-1[16, 17], cytokeratin (AE1/AE3) [18], myoepithelial marker p63 respectively [19]are used to rule out histological differential diagnoses.

It is remarkable that the tumor in our case with higher CD68 staining. Generally, the degree of inflammation measured by the expression of CD68 showed a positive significant correlation with tumor size and tumor growth index[20]. This is inflammatory in nature and may contribute to the pathogenesis of schwannoma[21]. Although there is a lack of clear insight regarding the control mechanisms for oncogenesis. In our case, the tumor with higher CD68 staining were likely to demonstrate that the tumor volume increase is not only based on cell proliferation, but also intratumoral hemorrhage, vascularization, and inflammation, which may be produce rarefaction of the bone and lead to bone fracture after a trivial trauma.

Ki-67 is a monoclonal antibody that provides a means of evaluating the growth fraction of normal and neoplastic human cell populations. A Ki-67 index of less than 3 per cent is expected for a typical schwannoma. schwannomas with an index of greater than 3 percent are presumed to be actively proliferating and pose a the oretically higher risk for regrowth or recurrence[22]. However, Ki-67 proliferative index is variable. In a study by Imagama *et al.*, Ki-67 index was < 1% in purely intramedullary tumors while it ranged between 18% and 25% in cases where the tumor was both intra and extramedullary[23]. In our case, the tumor was both intra and extramedullary. The ki-67 index was about 10%, which are presumed to be non-actively proliferating and pose a the lower risk for recurrence.

Finally, the recommended treatment for intraosseous neurilemmomais with pathologic fracture include excisional biopsy, curettage, bone allograft, and fracture fixation. In our case, there was no evidence of recurrence and have a good functional outcome.

Declarations

Funding None.

Data availability Data sharing not applicable to this article as no datasets were generated or analyzed during the current study.

Compliance with ethical standards

Conflicts of interest The authors declare that they have no conflict of interest.

Ethical approval No human or animal participants.

Informed consent Informed consent was obtained from all individual participants included in the study.

Author contributions All authors were involved in the conception and design of this article. Jiang Huajun and Yang Jingjing wrote the initial version of the manuscript; Qu Wei and Wu Yuxuan updated the manuscript.

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Figures

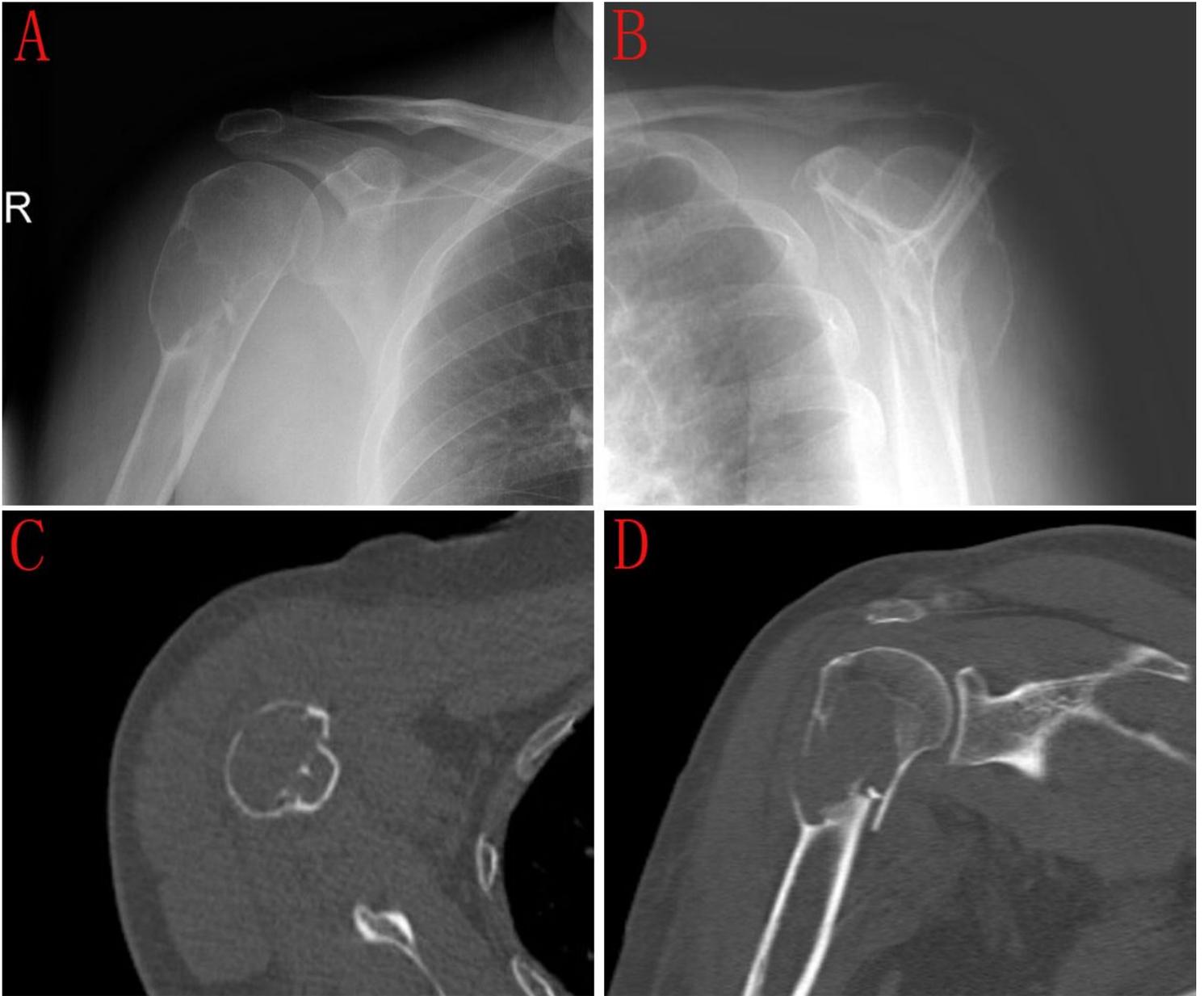


Figure 1

Anteroposterior (A) and lateral (B) radiographs, shows an expanding, lytic lesion affecting the proximal humerus associated with scalloping of the endosteum. Computed tomography (CT) Axial slice bone window (C) shows a intramedullary mass elevating the periosteum. Coronal slice bone window (D) shows a fracture at the proximal humerus region.

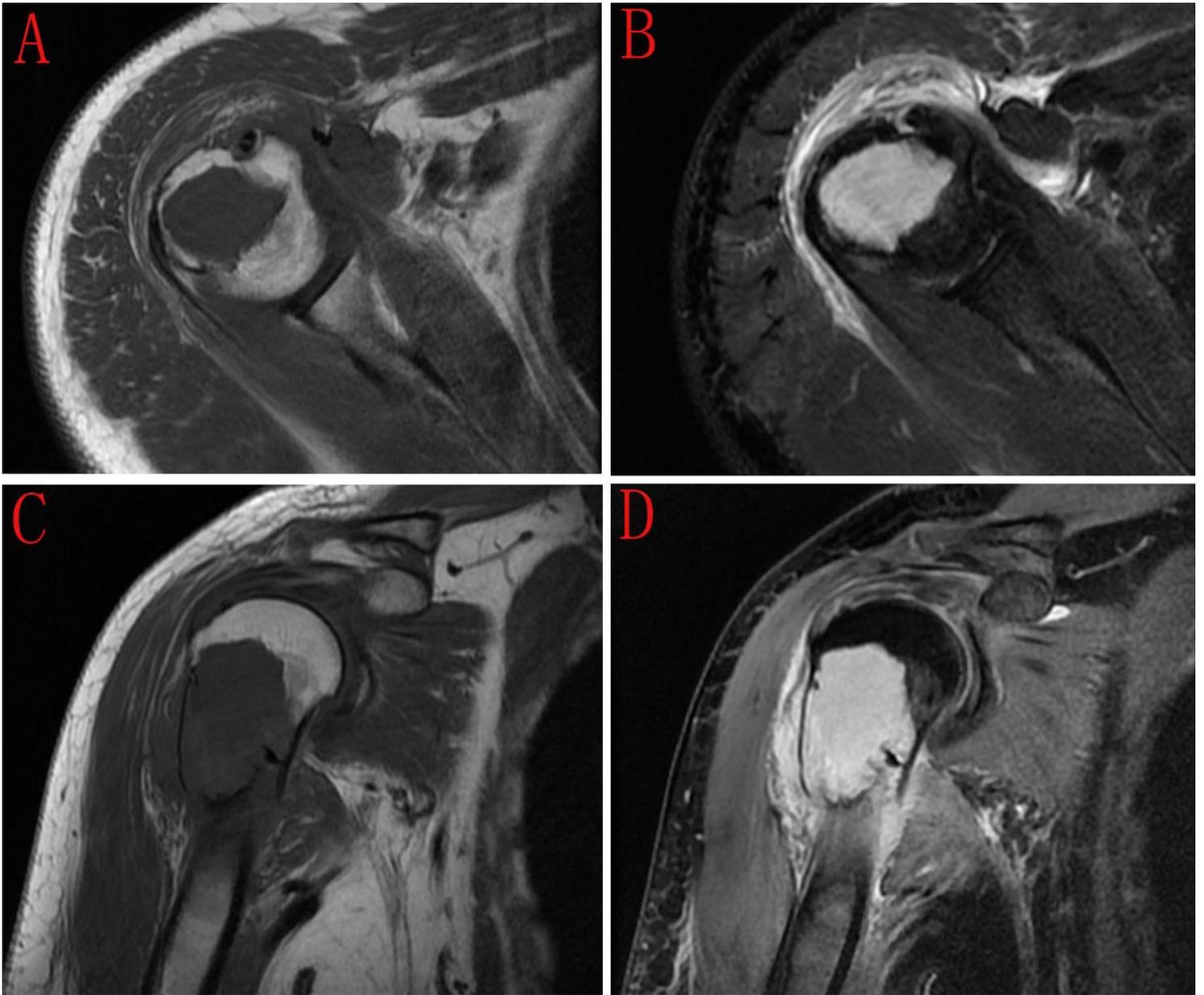


Figure 2

T1-weighted image shows an intramedullary lesion, isointense to muscle, (A) axial image, (C) coronal image. T2-weighted image shows a hyperintense lesion associated with soft tissue edema, (B) axial image, (D) coronal image.

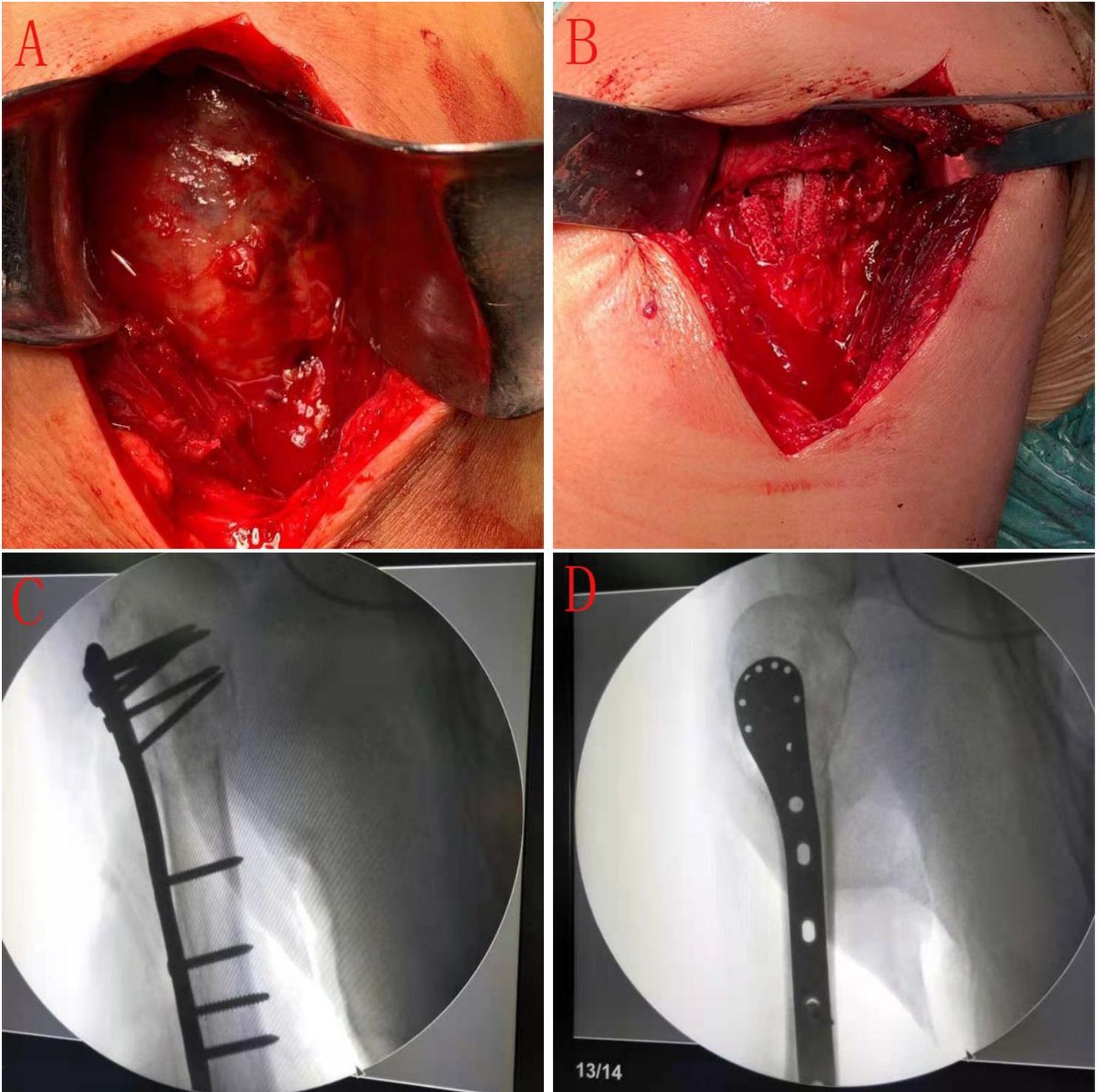


Figure 3

(A) Intraoperative , the tumor was a well-encapsulated soft grayish-yellow colored mass. (B) the tumor was removed and the cavity was filled with allograft bone. (C), (D) the pathologic fracture was fixated with a proximal humerus locking plate.

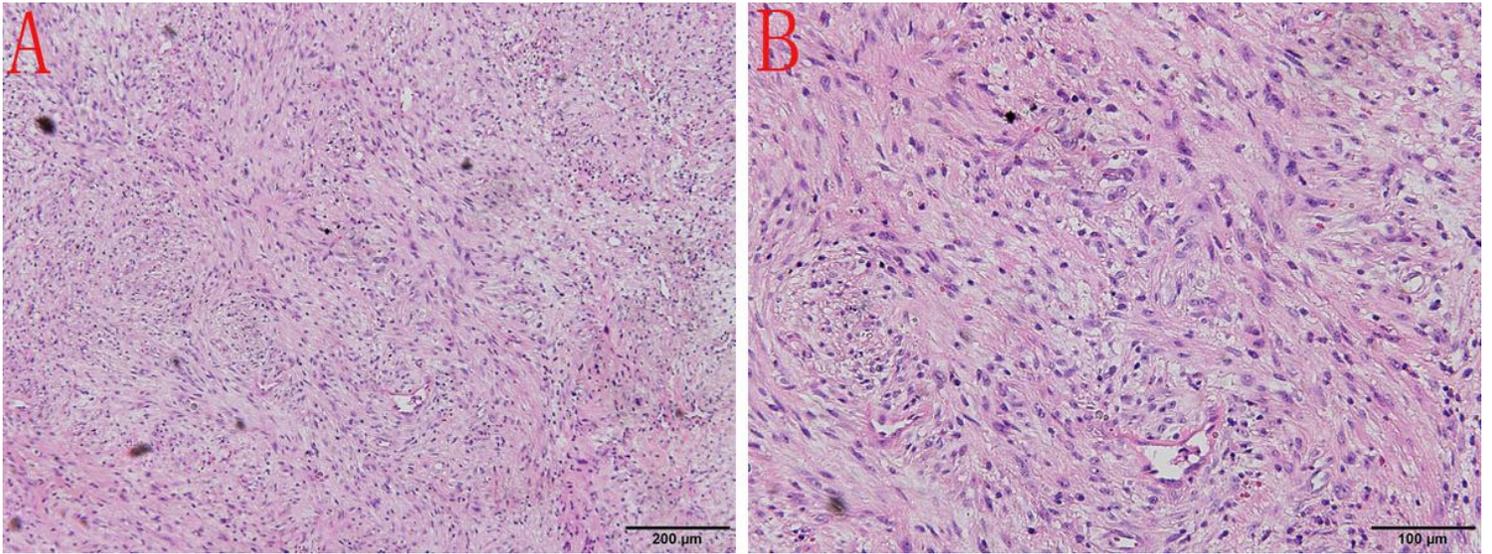


Figure 4

Pathological examination demonstrated typical picture of a schwannoma, showing whorls and interlacing fascicles of schwannoma spindle cells. (A) Hematoxylin and eosin staining,×100; (B) Hematoxylin and eosin staining,×200.

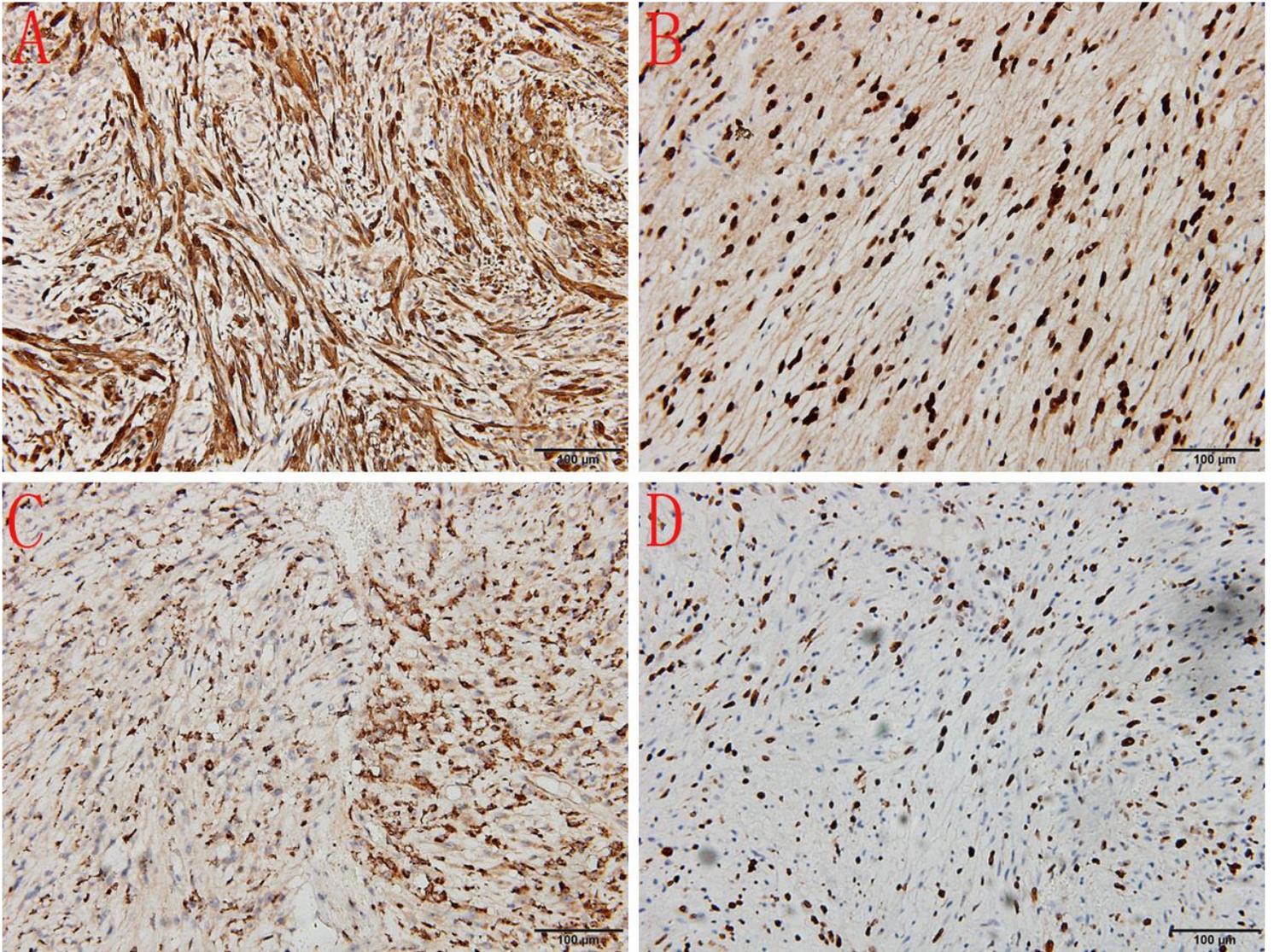


Figure 5

Immunohistochemical analysis demonstrated the presence of (A) S-100, (B) SOX-100, (C) CD68 and (D) Ki-67(+10%).

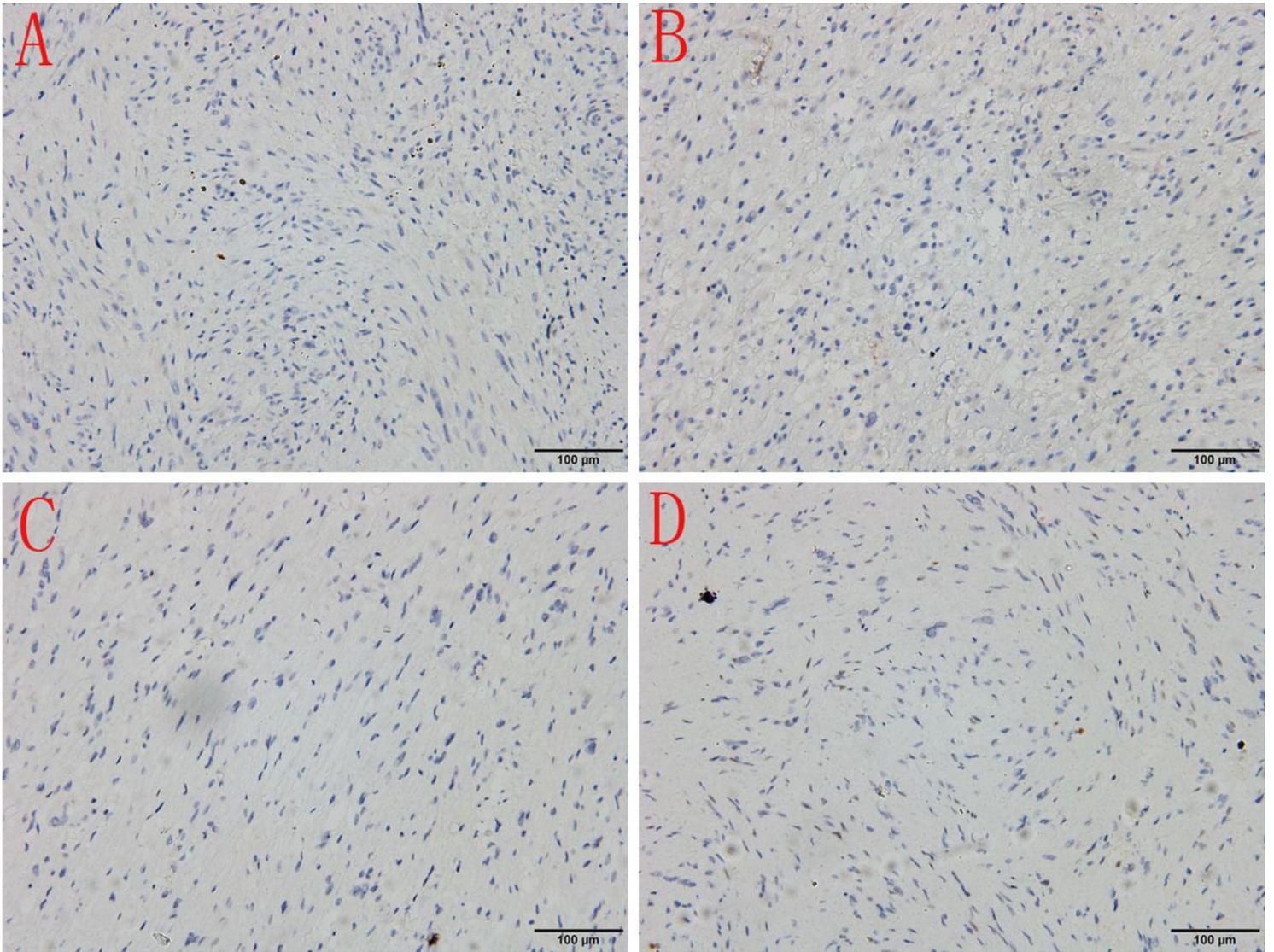


Figure 6

Immunohistochemical analysis demonstrated the absence of (A) Desmin, (B) DOG-1, (C) AE1/AE3, (D) P63.