Huge and deeply situated glomus tumor: Its malignant form

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Abstract

We report 3 cases of atypical glomus tumors which were suspected to be malignancy.

The most glomus tumors are small and single site that occur in the subungual region of fingers. However, all cases of this study were large size (>5cm) and deep location. They had no classic symptom of glomus tumor such as pain and tenderness. Affected sites were respectively the buttock, the ankle and the upper arm. One of the cases showed multiple lesions with cystic changes in the left leg.

It was reported that the metastatic rates of glomus tumors which size more than 2 cm and deep location was 67 %. Since all three cases of this study were deep-seated and more than 2 cm, they had malignant potential clinically. Therefore, future careful progress observation is required.

Key Wards　Atypical glomus tumors・Large size・Deep location

Introduction

Glomus tumors are rare, accounting for less than 2% of soft tissue tumors. They are most common in the upper extremities, particularly in the subungual region.[1] Glomus tumors are typically small (<1 cm), red-blue nodules that are often associated with a long history of pain, particularly with exposure to cold or minor tactile stimulation. However, occasional glomus tumors may show unusual clinical features, such as large size, deep soft tissue or visceral locations, infiltrative growth pattern, or multicentricity.[2-4] Very rare examples of glomus tumor have been considered to be histologically malignant.[5-7] The purpose of this paper is to describe three cases of atypical glomus tumors were suspected to be malignancy clinically, which are not histologically malignant.

Case reports

Case 1

A 34-year-old female presented with the chief complaint growing masses around the ankle. The mass was in her childhood and has been diagnosed as hemangioma at nearby hospital. Physical examination revealed an elastic hard mass, measuring 13×14 cm, on the medial side of the right ankle (Fig. 1a). There was mild tenderness and local heat. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed cystic masses around the right ankle

and the tumor lesions were not only medial side of the right ankle but also lateral and posterior side. MRI (Fig. 1b) showed low-to-iso signal intensity on T1-weighted images. On T2-weighted images, the mass had high signal intensity. The cyst walls were well-enhanced with contrast medium (gadolinium diethylene-triamine-pentaacetic acid [Gd-DTPA]). We suspected the tumor was synovial sarcoma or pigmented villonodular synovitis. An incisional biopsy was performed. Histological examination revealed small, uniform, rounded cells with a centrally placed, round nucleus and amphophilic to lightly eoginophilic cytoplasm (Fig. 1c). Immunohistochemically, the tumor cells stained highly positive for α-smooth muscle actin (SMA) and desmin was focally positive. CD34 and S100 protein were negative. A diagnosis of glomus tumor was made. We performed marginal excision of all tumors. All tumors were diagnosed as glomus tumors. The patient had no local recurrence and no lung metastasis at follow up 18 months after operation.

Case 2

A 22-year-old male had left buttock pain 5 years ago. He was referred to our institution after MRI showed a tumor lesion of the left buttock. Physical examination revealed an elastic hard mass, measuring 6×4 cm. In MRI (Fig. 2a), the tumor was shown slightly high signal intensity on T1-weighted images and high signal intensity on T2-weighted images. Principally, the

border of the tumor was well enhanced by Gd-DTPA. Because a malignant tumor, such as liposarcoma, was suspected from the physical and MRI findings described above, an open biopsy was performed. In the frozen section, diagnosis was something like malignancy. So, preoperative chemotherapy was started on the same day. But permanent histological findings were glomus tumors. We performed marginal excision after three weeks from the biopsy day. Final diagnosis was microscopically compatible with glomus tumor. The tumor cells are proliferating in dilated vascular wall and intravascular space with round nuclei and eosinophilic cytoplasm (Fig. 2b).

Case 3

A 73-year-old female experienced pain in her right upper arm that had persisted for 1 year. Physical examination revealed an elastic hard mass, measuring 4×5 cm, on the proximal side of the right upper arm. MRI (Fig. 3a) showed low signal intensity on T1-weighted images and high signal intensity, especially center area, on T2-weighted images. The wall of the tumor was enhanced after gadolinium injection. The tumor was revealed cystic mass. At operation, a mass four centimeters in diameter and reddish in color was noted. The mass showed no continuity with the shoulder joint. The tumor was removed in marginal margin. Microscopically, the tumors consisted of hemangiopericytomatou vascular features and were

composed of solid sheets cells with a round to oval nucleus. (Fig. 3b and c). Immunohistochemically, the tumor cells expressed SMA strongly. Desmin, CD34 and BCL2 were negative. The tumor was diagnosed as glomus tumor. The patient had no recurrence at 16 months after surgery.

Discussion

The first report of a clinically atypical infiltrating glomus tumor is the 1972 case report of Lumley and Stansfeld.[4] Atypical glomus tumors are admittedly rare and comprise less than 5% of glomus tumors.[8] The most glomus tumors are small and single site that occur in the subungual region of fingers. Occasionally, bony changes may arise due to glomus tumor.[9,10] The glomus tumors present a classic triad of symptoms: paroxysmal excruciation pain, point tenderness, and hypersensitive to temperature, especially coldness. None of these symptoms were observed in the present cases. While glomus tumors are usually solitary, multiple glomus tumors have been described by several authors.11-14 The Case 1 had multiple lesions. Cases with multiple tumors comprise less than 10 %.[11] Multiple glomus tumors are usually asymptomatic. The lack of classic symptoms triad may be a factor in the delay in diagnosis. [12]

Histologically, the glomus cells appear round to cuboidal with round/oval punched-out nuclei and slightly eosinophilic cytoplasm. Depending on the size of the nests of glomus cells,

the tumor may have a vascular appearance reminiscent of a hemangiopericytoma as in Case 3. However, a hemangiopericytoma can be distinguished from a glomus tumor because the pericytes in the glomus tumor have an organoid pattern, and because it is circumscribed, does not infiltrate and is always benign.[15]

Glomus tumors are usually small less than 1 cm in diameter and solid tumor. The sizes of the tumors in all present cases were more than 5 cm (Table 1) and Case 1 and 3 had cystic changes. Tachibana et al. reported cystic change of glomus tumor was very rare case.[16]

Folpe et al. described 52 cases of atypical and malignant glomus tumors.[8] They reported the metastatic rates of glomus tumors which size more than 2 cm and deep location was 67 %. According to their diagnostic criteria, malignant glomus tumor fulfilled at least one of following: the tumor with a deep location and more than 2 cm, or presence of atypical mitotic figures, or a combination of moderate to high nuclear grade and mitotic activity (5 mitoses/50 high-power fields). According to World Health Organization (WHO) classification of tumors, there are two types of histological malignant glomus tumor. In the first type, the malignant component retains overall architectural similarity to benign glomus tumor and consists of sheets of round cells such as Ewing sarcoma. In the second type, the malignant component resembles a leiomyosarcoma or fibrosarcoma. As for the present cases, the all lesions were more than 5 cm and deep location, but the histological malignant futures were not detected.

The tumors were malignant glomus tumors according to Folpe's diagnostic criteria. Because the metastasis tends to appear long after surgery, close long-term postoperative follow-up is necessary for the present cases.

In conclusion, we described three cases of atypical glomus tumors that malignant glomus tumors were suspected, because all tumors deeply located and were more than 5 cm in size. Therefore, work-up for metastases and local recurrences should be taken in consideration.

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Figure legends

Fig. 1, Case 1

a. Clinical photography. A huge mass is shown in her right ankle.

b. Coronal magnetic resonance (MR) images showing the masses on bilateral side which had low signal intensities on T1-weighted images, high signal intensities on T2-weighted images and enhancement of cyst wall (arrows) with gadolinium diethylene-triamine pentaacetic acid (Gd-DTPA).

c. Tumor cells were small, uniform, rounded shape with a centrally placed, round to oval nucleus and eoginophilic cytoplasm. (H&E ×200)

Fig. 2, Case 2

a. Axial MR images showing the mass on left buttock which had slightly higher intensities on T1-weighted images, high signal intensities on T2-weighted images and enhancement of marginal area (arrows) by Gd-DTPA.

b. The tumor cells are proliferating in dilated vascular wall and intravascular space. (H&E ×20)

Fig. 3, Case 3

a. Coronal MR images showing the tumors on right upper arm which had low signal intensities on T1-weighted images and high signal intensities on T2-weighted images. The

cyst wall was enhanced after gadolinium injection.

b. Microscopically, the tumors consisted of hemangiopericytomatou vascular features. (H&E ×20)

c. Most tumors were composed of solid sheets cells with a round to oval nucleus. (H&E ×200)

Table 1, Clinical features of malignant glomus tumors

Fig. 1-a

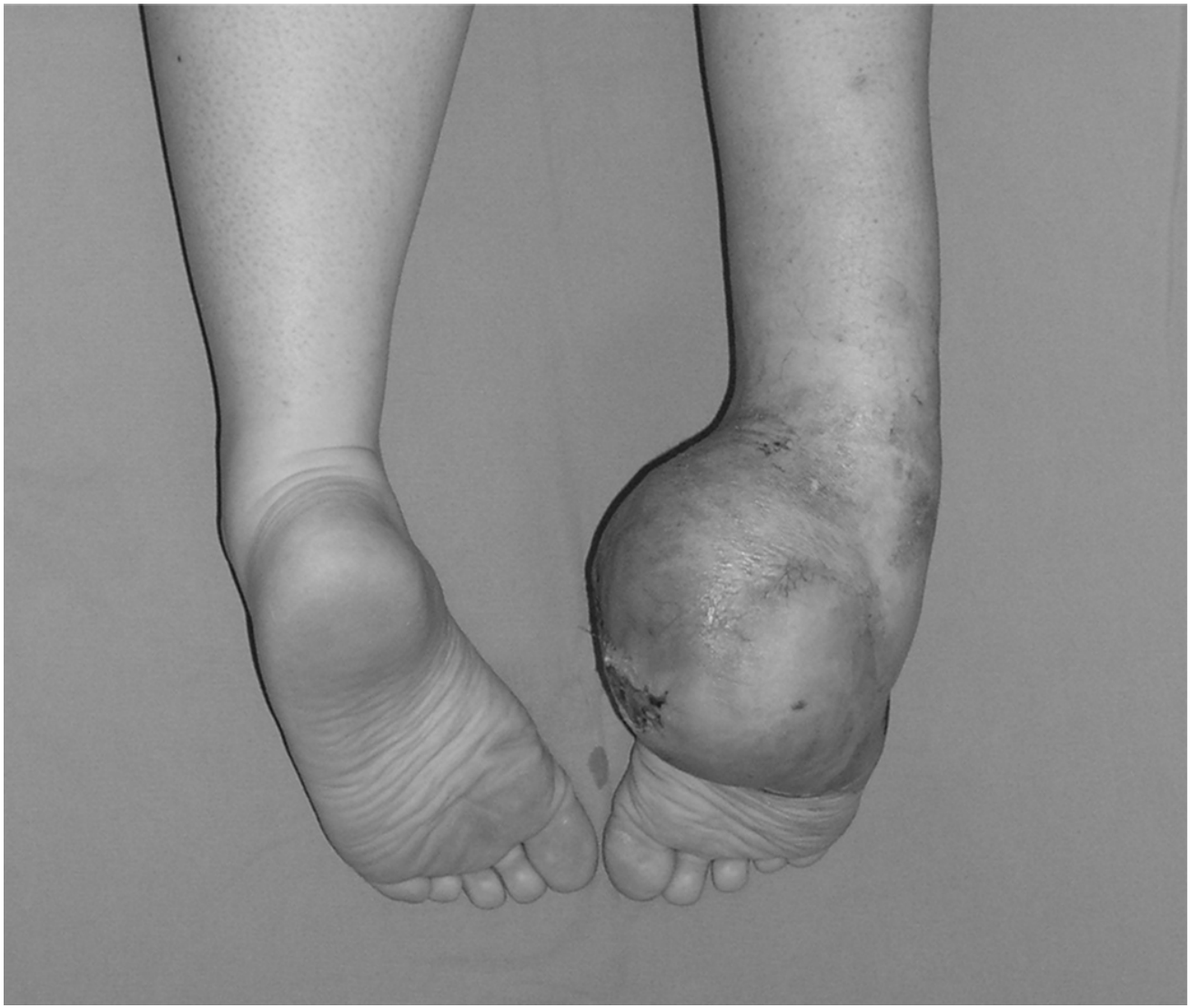

Fig. 1-b

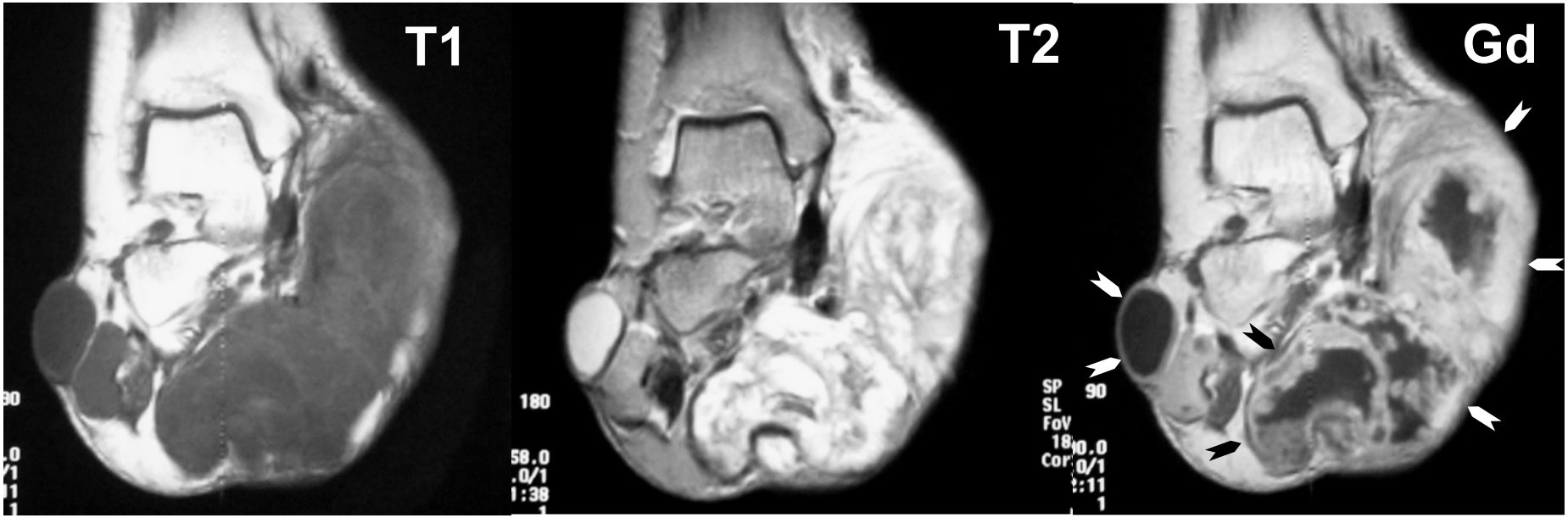

Fig. 1-c

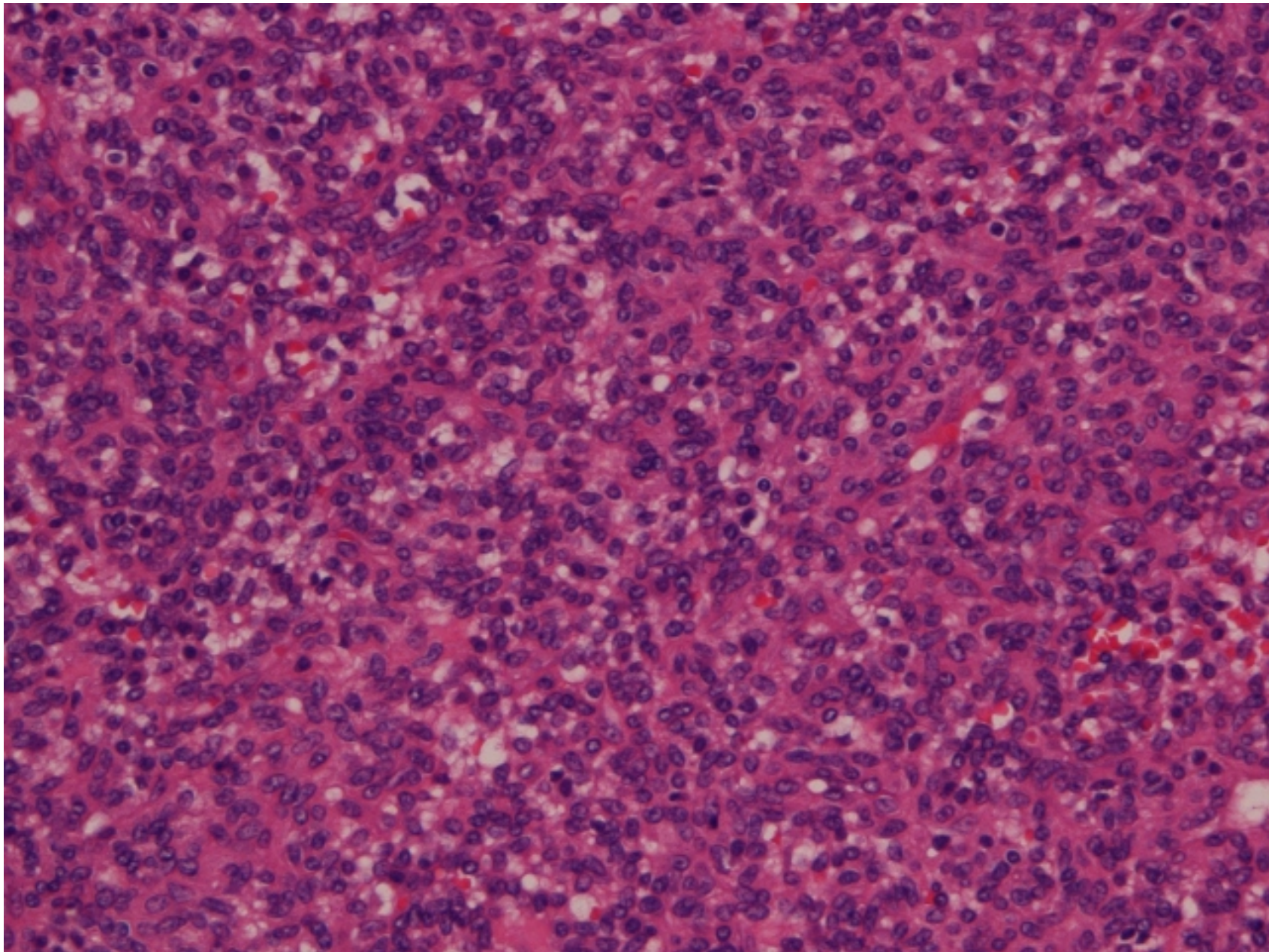

Fig. 2-a

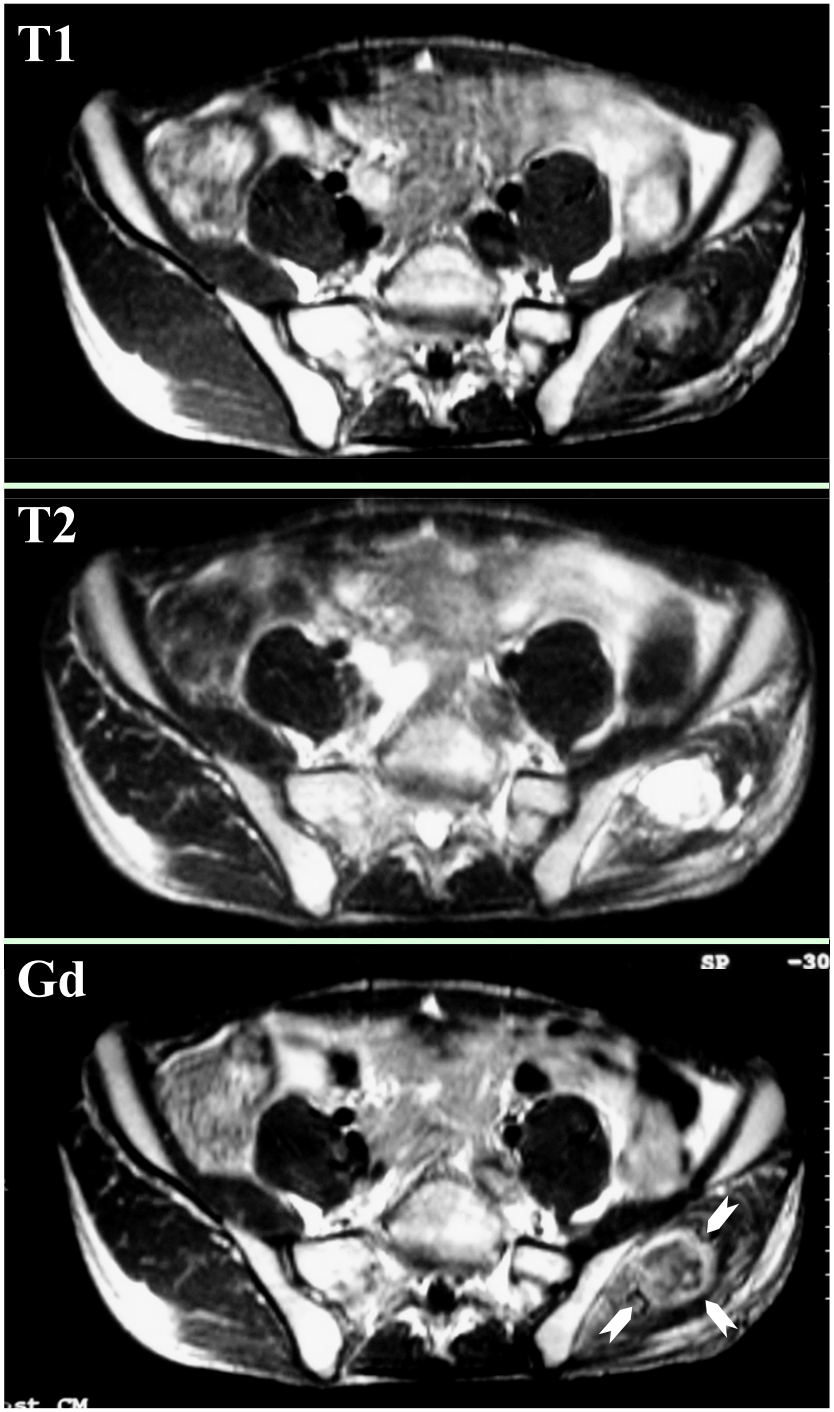

Fig. 2-b

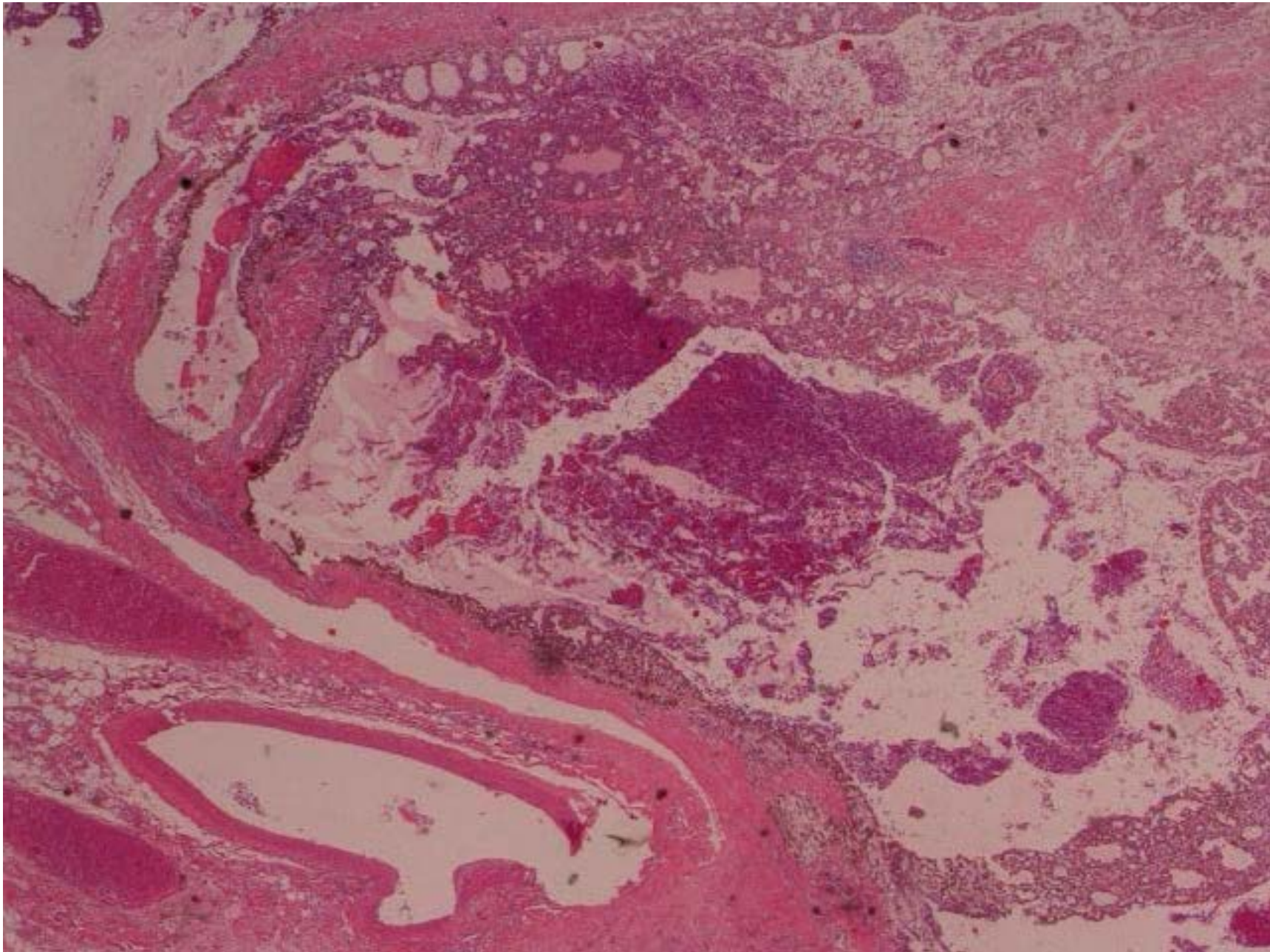

Fig. 3-a

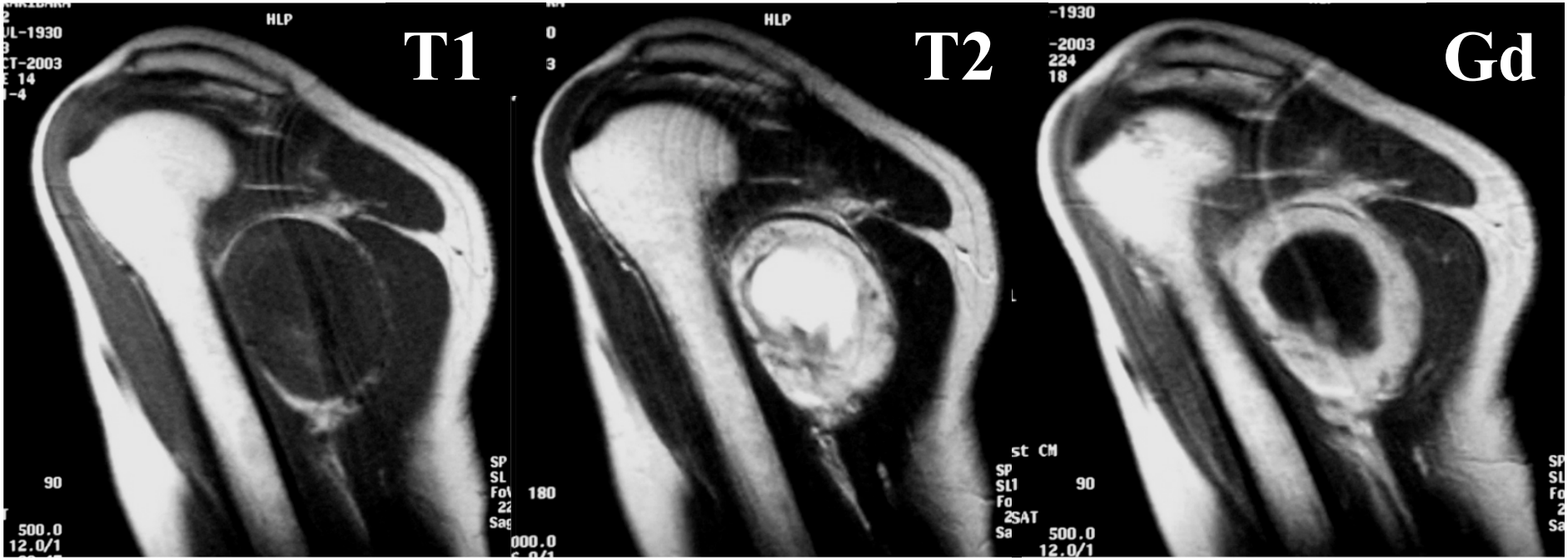

Fig. 3-b

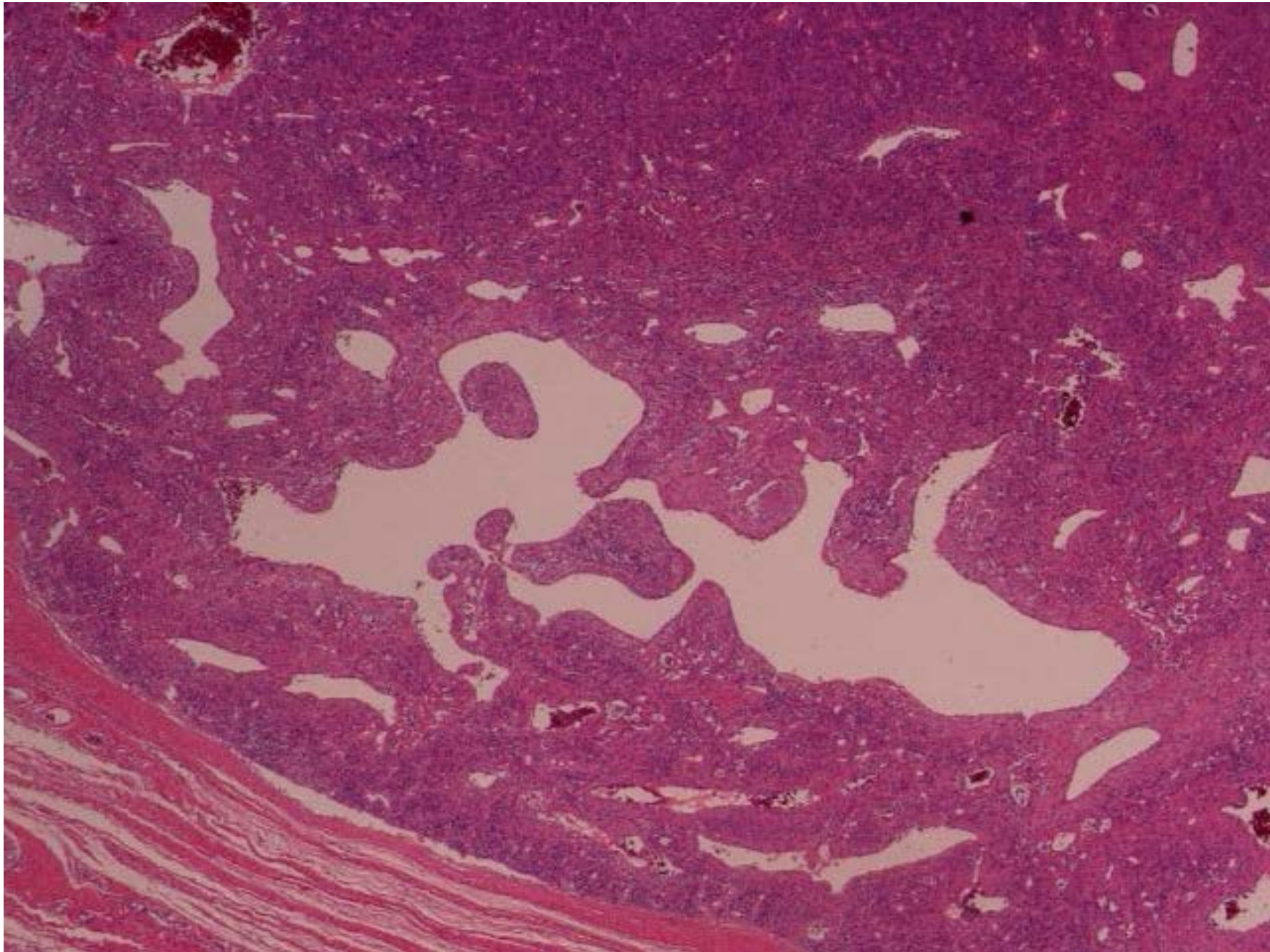

Fig. 3-c

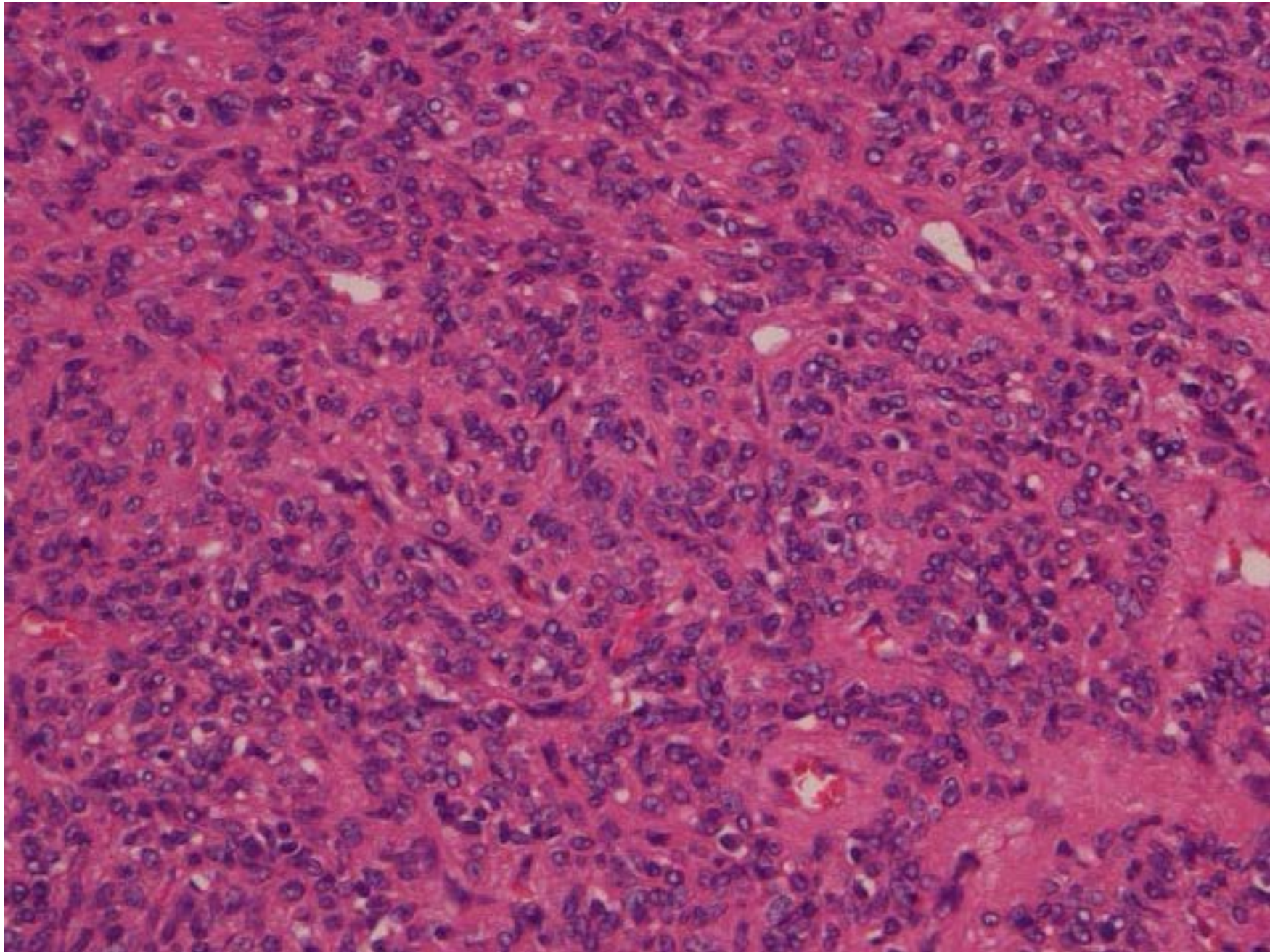

Table 1

	Age	Sex	Site	Size (cm)	Depth	Mitosis
Case 1	34	Female	Ankle	12×13	Deep	No
Case 2	22	Male	Buttock	6×4	Deep	No
Case 3	73	Female	Upper arm	5×4	Deep	No