One Case Report of Desmoplastic Small Round Cell Tumor of Pleura

Xiao-bo Zhang
Lanzhou University Second Hospital
Sheng-lin Li
lanzhou university senond hospital
Xin Su
Gansu Provincial Hospital
Rui-hao Zhang
Lanzhou University Second Hospital
Yi-cun Hu
lanzhou university senond hospital
Ke-Ping Wang
Lanzhou University Second Hospital
Hai-yu Zhou (zhouhy@lzu.edu.cn)
Lanzhou University Second Hospital

Case report

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Abstract

BACGROUND: Desmoplastic small round cell tumor (DSRCT) is a rare, aggressive malignancy tumor that often affects the abdomen, especially in males. Extra-abdominal DSRCT is extremely rare.

CASE REPORT: Here we report a 33-year-old man diagnosed with DSRCT from pleura without any clinical symptoms. Routine blood examination was normal, and an X-ray examination revealed a chest mass during the routine physical examination. The patient was diagnosed by immunohistochemistry combined with clinical features and imaging examination and finally confirmed by pathology. After the diagnosis, we immediately underwent surgery and postoperative radiotherapy. In the latest follow-up, there was no sign of recurrence.

CONCLUSION: Because the disease is asymptomatic at the beginning, it is easy to deteriorate, and there is no specific targeted treatment. Therefore, early diagnosis and treatment are particularly important.

1. Introduction

DSRCT is a rare, multifocal peritoneal malignancy with the frequently disseminated abdominal disease at presentation, which often occurs in young adults, especially in males. The disease most commonly involves the omentum and peritoneum, followed by the retroperitoneum [1]. Pleural DSRCT is a kind of extra-abdominal DSRCT, which is rare in the lung [2], paranasal sinuses [3], central nervous system [4], soft scalp tissue [5], salivary glands, and pleura [6]. The clinical manifestations are nonspecific and usually related to the size and location of the tumor. Among them, the translocation of T (11; 22) (P13; Q12) was regarded as its unique marker [6, 7]. The disease often signifies a poor prognosis, and anticancer treatment is required, including surgery, chemotherapy, radiotherapy. Combination therapy may prolong survival time [8, 9]. Here we report a case about pleural DSRCT.

2. Case Report

A 33-year-old man was found a mass in the thoracic cavity by chest X-ray during a routine physical examination. He never had any clinical symptoms, such as chest pains and breathing difficulties, with no smoking history. Chest CT underwent two weeks later showed there were space-occupying lesions in the right pleural and discontinuity of the fourth anterior rib in the right. Then he was admitted to the department of thoracic surgery. Physical examination reveals normal breath sounds. Tumor markers of lung cancer such as CEA, NES, CY211, SCC were in the normal range. Enhanced Chest CT with three-dimensional reconstruction revealed that the tumor appeared as a round-like, sharply defined lesion in the right thoracic cavity. It was about 4.7×4.2×4.0 cm and slightly hypodense, which was based on the pleura. There was a low attenuation (29 Hounsfield Units, HU) on plain examination and heterogeneous enhancement on contrast-enhanced CT, both arterial (47 HU) and venous phases (63 HU) (Fig. 1). The tumor density was uniform, and there was no necrotic cystic change. The fourth ribs had localized bone destruction, which was slightly strengthened after enhancement. PET/CT illustrated a locally

hypermetabolic lesion in the right fourth anterior rib (Fig. 2). No abnormalities were found in the enhanced abdominal CT examination. Cranial MRI showed no apparent abnormalities. The patient underwent the Resection of Chest Wall Tumors and a costectomy. The patient was treated by radiotherapy on his second visit. There was no metastasis and recurrence of the tumor. Besides, our team will continue to follow up on patients' tumor metastasis and prognosis.

3. Availability Of Data And Materials

DSRCT has no specific diagnostic criteria, and it mainly relies on multiple positive immunohistochemical stains to diagnose the disease. Microscopically, the size of the small tumor cells was consistent, the nucleus was small and deeply stained, and the cytoplasm was sparse, the boundary was not clear, arranged in nests, surrounding sclerotic stroma (Fig. 3).

Immunohistochemical staining showed: partial cells CD57 (+), Bcl-2 (+), CD99 (+), EMA (+), viementin (+), Desmin (+), CKp (-), SMA (-), CK5/6 (-), p63 (-), Calponin (-), CD20 (-),CD3 (-), CD79a (-), mum-1 (-), λ (-), κ (-), CD38 (-), CD138 (-), CD30 (-), TdT (-), CD21 (-), S100 (-), CD34 (-), myoD1 (-), myogenin (-), HMB45 (-), MelanA (-), Syn (-), CgA (-), TTF-1 (-), NSE (-), OCT34 (-), SALL-4 (-), STST6 (-), CK8/18 (-), 15% Ki67-positive cells (Fig. 4). This is similar to the previous results reported by immunohistochemistry [10]. The diagnosis of DSRCT of the pleura was eventually established with no rib invaded.

4. Discussion

The 5-year survival rate of DSRCT is only 15%. Pathological examinations usually show cell nests with clear outlines in different shapes and sizes and obvious sclerotic stroma around the nests [11]. Histopathological examination can help give a diagnosis [9]. Pleural DSRCT is extremely rare. The previous review about pleural DSRCT studied fifteen patients, of which the mean age is 25.5 and male-dominated. All patients presented chest or back pain and respiratory symptoms, including dyspnea, cough, pleural effusion. The patient we report didn’t have any clinical symptoms though the abnormality was reflected by a chest X-ray. Due to a lack of special clinical manifestations, clinicians may ignore the disease. Histology biopsy is quite important [12]. 10 of 15 patients showed Desmin (+), 4 EMA (+), 4 CD99 (+). The patient we report showed CD57 (+), Bcl-2 (+), CD99 (+), EMA (+), vimentin (+), Desmin (+).

Early diagnosis is difficult because most tumors show nonspecific gastrointestinal symptoms after a long asymptomatic period [13]. Treatments involve surgery followed by adjuvant radiotherapy or chemoradiotherapy. The survival period reported range from 4 months to 76 months [6]. Combination therapy may prolong the survival time of patients. But it grows rapidly, has a high recurrence rate, and has a poor prognosis, the five-year overall survival rate is 15% [13]. Therefore, targeted therapies are urgently needed.

5. Conclusions

Clinicians can't ignore the disease though there are no clinical symptoms.

Abbreviations

DSRCT: Desmoplastic small round cell tumor

HU: Hounsfield Units

CT: Computed Tomography

MRI: Magnetic Resonance Imaging

Declarations

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Author contributions

Resources: Sheng-lin Li

Supervision: Ke-ping Wang, Hai-yu Zhou

Writing – original draft: Xiao-bo Zhang, Xin-Su

Writing – review & editing: Rui-hao Zhang, Yi-cun Hu

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Availability of Data and Materials

All data generated or analyzed during this study are included in this published article.

Ethics approval and consent to participate

Not applicable.

Consent for publication

Written informed consent for publication was obtained from the participant.

Conflict of interest

The authors declare that there are no conflicts of interest.

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Figures

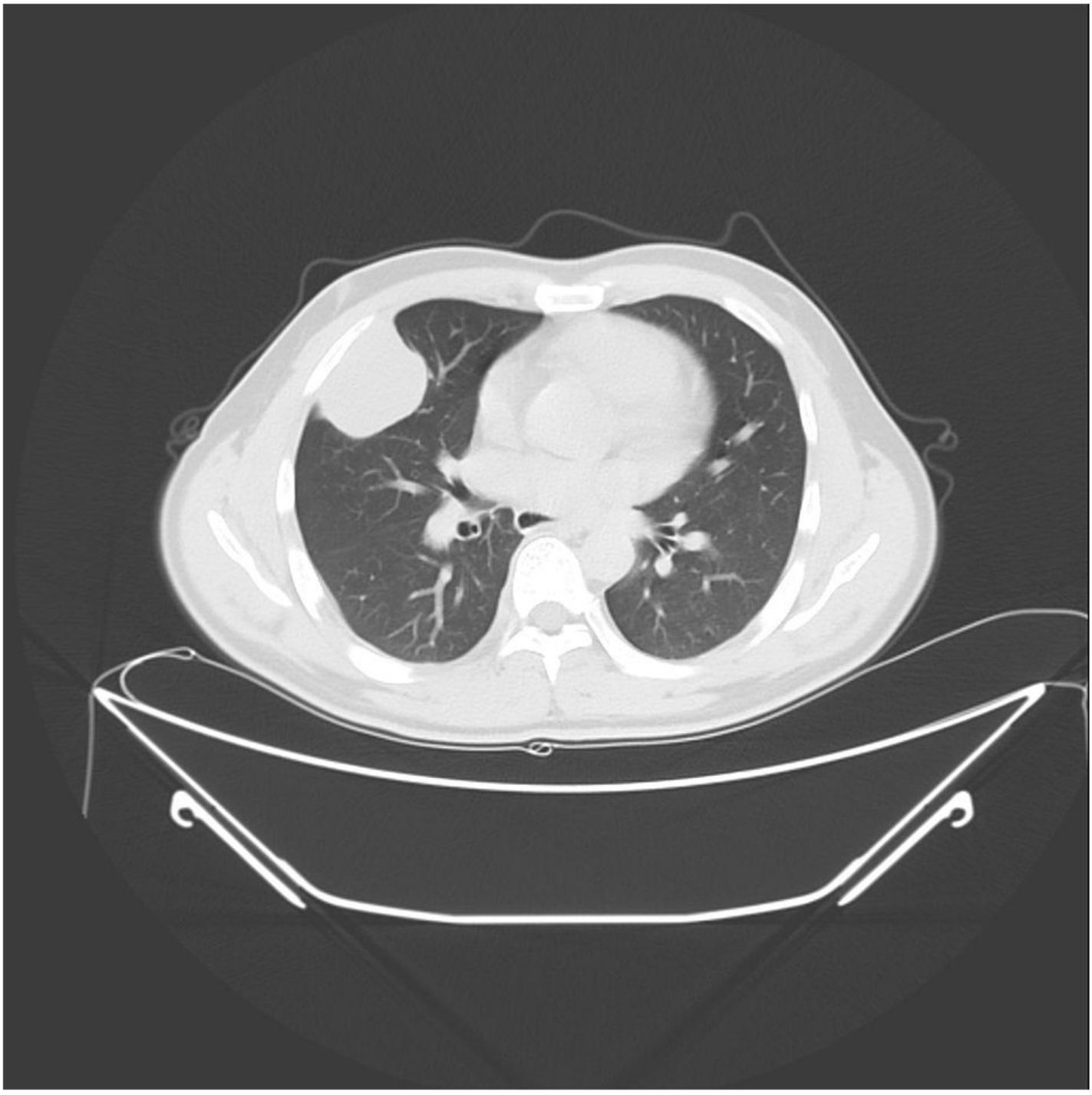

Figure 1

Pulmonary Window in enhanced CT showed the round mass is based on the pleura.

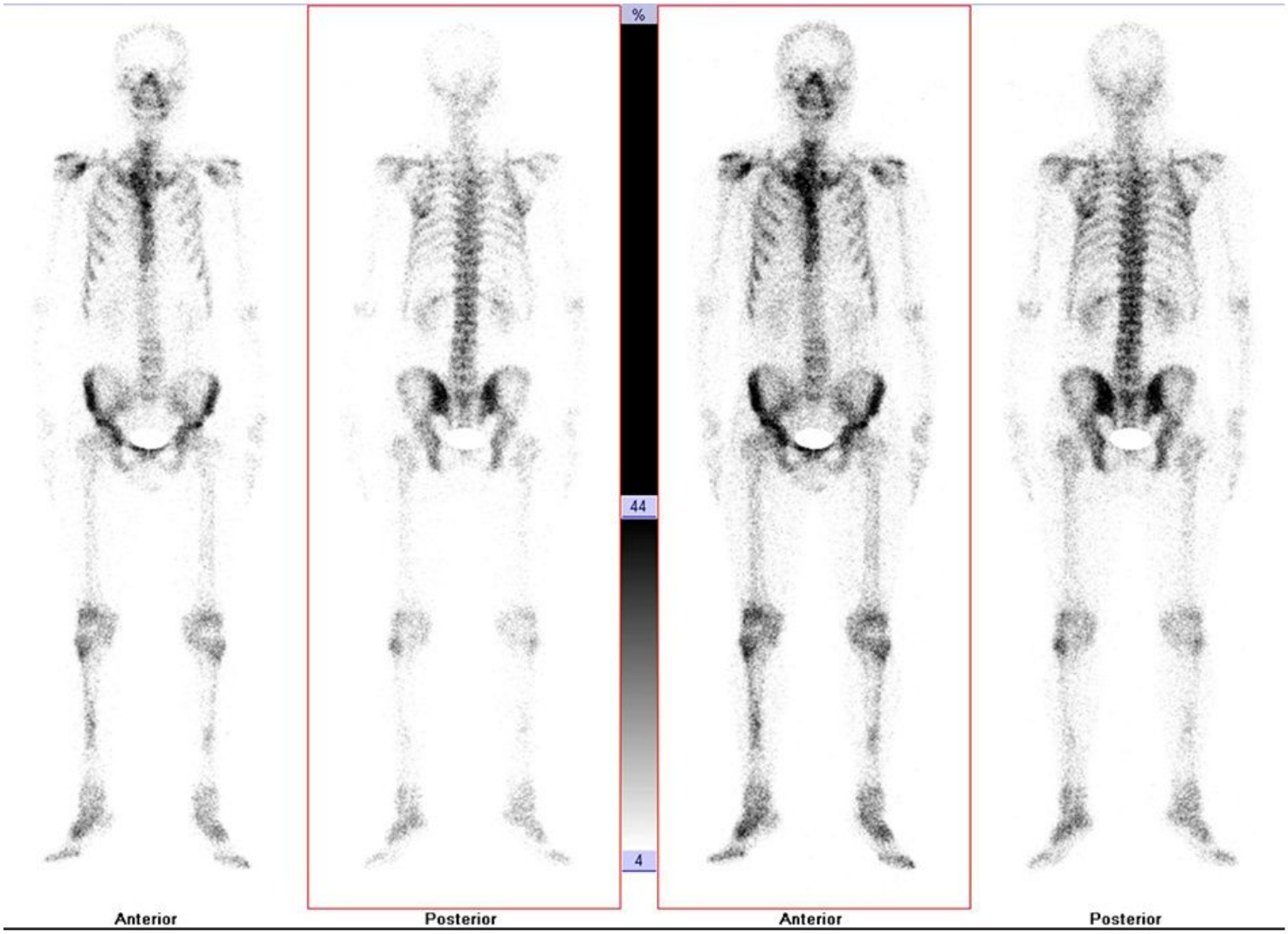

Figure 2

PET/CT illustrated a locally hypermetabolic lesion in the right fourth anterior rib.

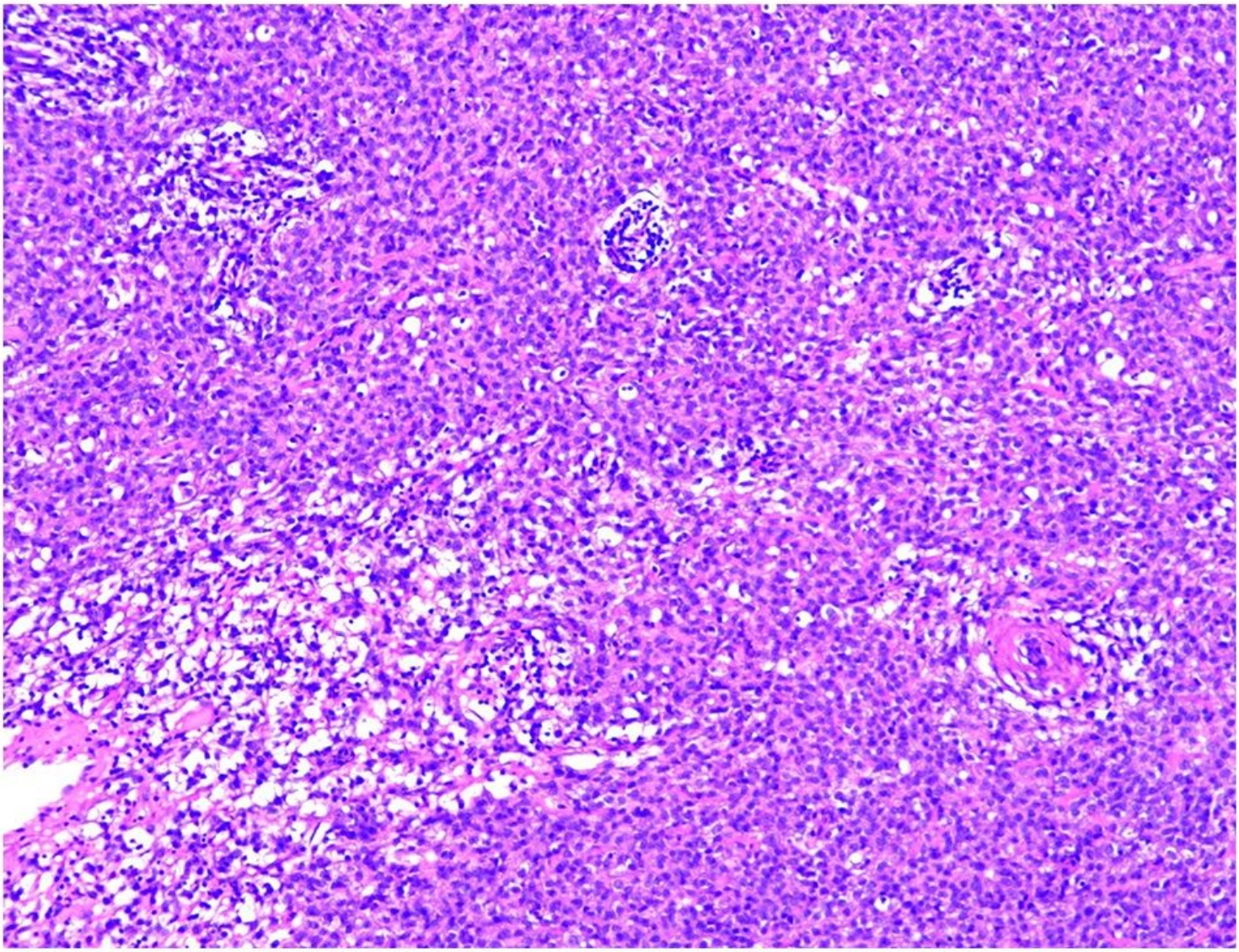

Figure 3

Tumor cells are arranged in nests with sclerotic stroma around them.

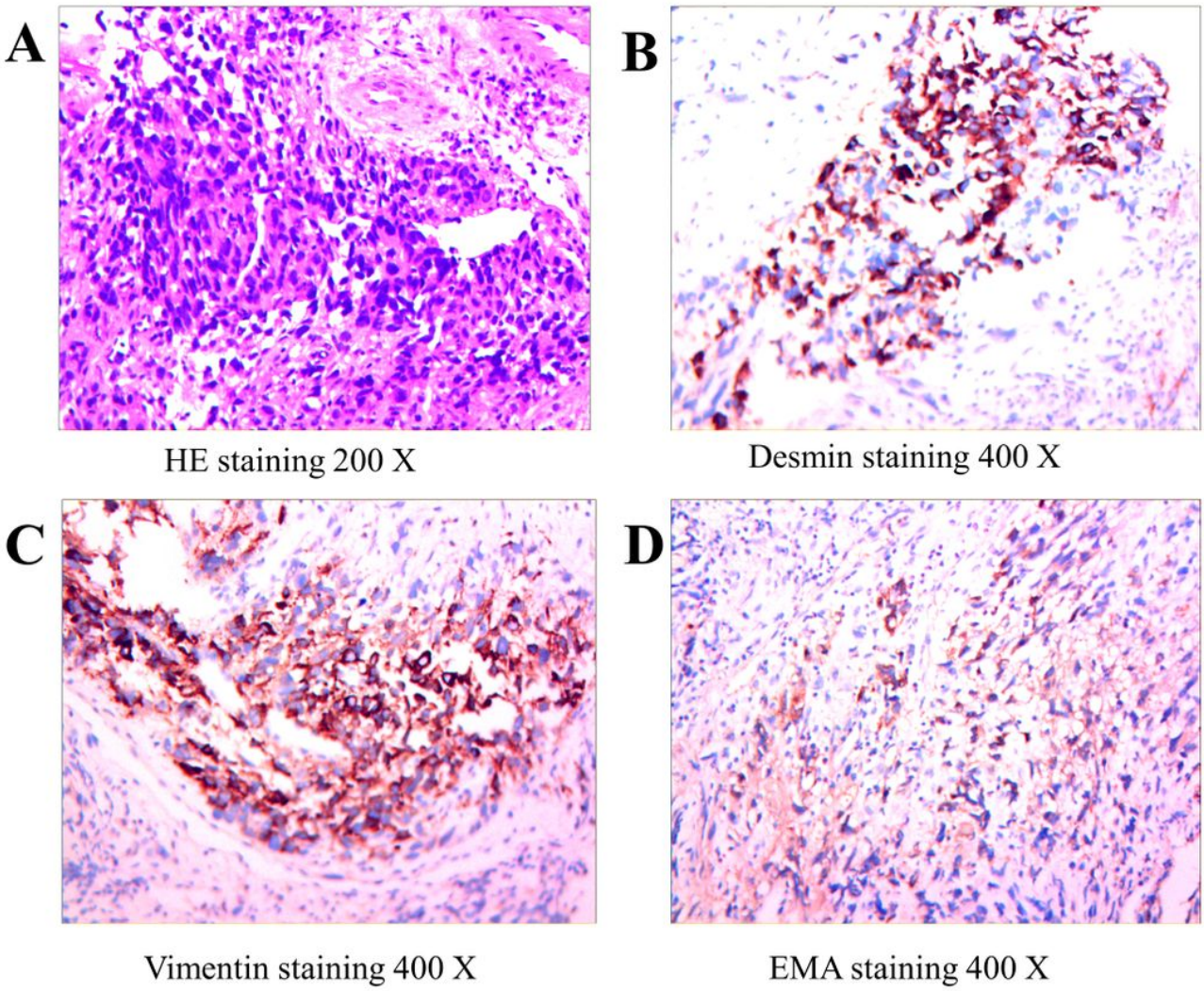

Figure 4

Immunohistochemical staining.