Hodgkin's disease of the thoracic vertebrae

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Abstract

Background Context: Hodgkin's disease rarely occurs in the spine, which is usually a setting for the advanced form of the disease.

Purpose: To describe an unusual case of isolated, primary spinal Hodgkin's disease, and to draw attention to this disease as a possible diagnosis in patients with mixed inflammatory cell infiltrate lesions located in the thoracic spine.

Study Design/Setting: A case report of a 28-year-old woman who presented with back pain and progressive weakness in the lower extremities as a result of spinal cord compression due to Hodgkin's disease of the thoracic vertebrae.

Methods: We report a new case of spinal cord compression due to Hodgkin's disease of the thoracic vertebrae. Decompression surgery was performed in the patient, followed by antibiotic treatment.

Results: Antibiotic therapy temporarily improved inflammation and fever. However, magnetic resonance imaging (MRI) evaluation showed that the inflammatory reaction in the lesion was not completely resolved. The disease progressed and later investigations

revealed Hodgkin's disease, which improved with a course of chemotherapy and radiation.

Conclusions: Hodgkin's disease should be considered in the differential diagnosis of spinal

neoplastic lesions with clinical features similar to spondylitis. Since MRI evaluation

showed that the vertebral disc was maintained in this case, the presence of a tumor rather

than inflammation should have been suspected.

INTRODUCTION

Hodgkin's disease most commonly presents with progressive but painless enlargement of peripheral lymph nodes, especially around the cervical region. Non-Hodgkin's lymphoma arises in the lymph nodes in approximately 60% of patients and at extranodal sites in approximately 40% of patients [1]. On the other hand, Hodgkin's disease is believed to arise in the lymph nodes in 90% of patients, and at other sites in the remaining 10% of patients. The incidence of Hodgkin's disease with spinal cord compression is rare, and the spine is usually a setting for the advanced form of the disease [2]. An initial manifestation of spinal cord compression due to Hodgkin's disease in the spine is extremely rare. We describe a 28-year-old woman who initially presented with spinal cord compression due to Hodgkin's disease of the thoracic spine.

CASE REPORT

A 28-year-old woman presented with a history of upper back pain that began after January 2002, 5 months before admission. Her height was 156 cm, and her weight was 45 kg. A month before admission, she began to develop progressive weakness in the lower extremities

and numbness below the upper thoracic area. Her medical history indicated no abnormalities.

A physical examination on admission revealed remarkable bilateral lower extremity

weakness that was worst in the hip flexors but did not preclude walking. A subjective

decrease in pinprick sensation below the 4th thoracic dermatome was noted. Increased tone

and hyperreflexia in the lower extremities, with bilateral Babinski signs, were also observed.

A T2-weighted sagittal magnetic resonance imaging (MRI) scan of the thoracic spine

showed a high-signal mass in the region anterior to the 1st– 3rd thoracic vertebra and an

epidural lesion (Fig. 1). Extensive soft tissue swelling and enhancement around the lesion,

with kyphotic deformity and spinal cord compression, were present. Contrast-enhanced MRI

showed that the epidural space and spinous process around the vertebral level from the

1st–3rd thoracic vertebra were enhanced, but the lesion did not invade the intervertebral disc.

Routine blood analysis gave the following results: white blood cell count, 9900/μL;

C-reactive protein level, 6.24 mg/dL; erythrocyte sedimentation rate, 114/118 mm; soluble

interleukin (IL)-2 receptor level, 865 U/mL; and normocytic anemia (red blood cell count,

373×10^4/μL; hemoglobin, 10.4 g/dL; hematocrit, 32.0%). In addition, the results of various

tests for infection and a tuberculin skin test were negative. Analysis of chest radiographs

indicated no abnormalities besides the destructive thoracic spine lesion associated with a soft

tissue mass and kyphotic deformity. Computed tomography (CT) demonstrated osteolytic

change of vertebral bodies, pedicles, and laminas in both the 2nd and 3rd thoracic vertebrae.

In particular, a lesion near the muscle density presenting at the site of the vertebral body

from anterior to lateral in the 2nd thoracic vertebra and tracheal compression were seen.

Gallium-67 scintigraphy showed abnormal accumulation around the 2nd thoracic vertebra.

Considering the above-mentioned diagnostic information, the differential diagnosis included

spinal tuberculosis, pyogenic spondylitis, malignant lymphoma, and other neoplastic lesions.

Since the paralysis of both legs became worse and MRI showed an increase in lesion size 3

weeks after admission, we performed posterior decompression from the 1st–3rd thoracic

vertebra, open biopsy, debridement, and drainage. Histological findings from the tissue

removed during surgery showed chronic inflammatory infiltrates with necrosis, but no

neoplastic cells. Bacterial and fungal cultures were negative. We performed polymerase

chain reactions with the tissue, but tuberculosis, *Mycobacterium avium*, and *Mycobacterium*

intracellulare tests were negative. Acid-fast bacilli were detected (Gaffky 2). Antibiotic

treatment with new quinolones and macrolides improved the inflammatory reaction and

clinical manifestation, and the spine lesion became smaller. Therefore, we assumed that the

lesion was spondylitis due to an atypical mycobacterium, and performed chemotherapy with

new quinolones, macrolides, and antituberculous medications.

However, the inflammation gradually worsened, with a 2- or 3-week cycle of remission and exacerbation. Thus, we concluded that antibiotic treatment could not improve the inflammation and performed a posterior-approach anterior spinal cord decompression from the 1st–3rd thoracic vertebra, open biopsy, debridement, and drainage 5 months after admission. Histological analysis showed no neoplastic cells. An acid-fast bacterial stain was also negative, and there was only inflammatory infiltration present. Even after surgery, the inflammatory reaction did not improve.

Six months after admission, chest CT showed a large lesion at the anterior mediastinum, and CT-guided needle biopsy was performed at the site (Fig. 2). Histological analysis (hematoxylin-eosin [H.E.] ×10) of the biopsy specimen showed inflammatory infiltrates with fibrosis and necrosis (Fig. 3). Reed-Sternberg (RS) cells were seen under a high-power field (×200), as well as lymphoma cells that were immunoreactive for CD15 and CD30 (Fig. 4). At this point, we diagnosed spinal cord compression due to Hodgkin's disease of the thoracic vertebrae. The antibiotic and antituberculous medications were stopped, and the patient was treated with adriamycin, bleomycin, vinblastine, and decarbazine (ABVD) and subjected to radiation therapy of the thoracic spine. The lesion markedly reduced in size. The

inflammatory reaction and clinical condition were also significantly improved. Due to the

reduction in lesion size and clinical improvement observed after 12 cycles of ABVD, we

performed posterior fusion from the 5th cervical to the 7th thoracic vertebra 13 months after

admission, because we had performed bilateral total facetectomy during posterior-approach

anterior spinal cord decompression previously. When we performed posterior-approach

anterior spinal cord decompression, we could not use instrumentation because we were wary

of infection. The patient's lower extremities showed improved strength and she was able to

return to work. MRI at the 6 years after surgery showed no evidence of recurrence (Fig. 5).

No further evidence of disease has been noted elsewhere in the patient during 10 years of

follow up (Fig. 6).

DISCUSSION

Hodgkin's disease accounts for 0.5–1% of all adult neoplastic diseases [3]. In 90% of

patients with Hodgkin's disease, the disease is thought to arise in the lymph nodes, whereas

in the remaining 10% of patients the disease arises at other sites [1]. Spinal lesions occur in

5.8% of Hodgkin's disease patients [4]. Wood and Coltman estimated that primary

extranodal presentation of Hodgkin's disease occurs in < 0.25% of patients [5]. Spinal lesion with extranodal Hodgkin's disease as the primary presentation is even more uncommon.

Spinal cord compression caused by a malignant lymphoma is generally a late manifestation of the disease [6]. There are about 3 times more patients with non-Hodgkin's disease than those with Hodgkin's disease [7]. Between 0.1–6.5% of patients with non-Hodgkin's lymphoma develop spinal cord compression at some time during the course of their disease, usually because of an epidural tumor mass. Fifty of 1039 patients (5%) with non-Hodgkin's lymphoma in one series developed central nervous system (CNS) involvement [8]. Four percent had leptomeningeal disease, 1% had intracerebral lesions, and 0.1% had spinal cord compression. Only 5 of the 50 patients (0.5%) had CNS involvement at the time of initial presentation. In another series, 9% of patients with non-Hodgkin's lymphoma developed CNS involvement [9]. The sites involved were the leptomeninges (4%), the spinal epidural space (3.5%), and within the cerebrum (1.5%). Half of the cases of spinal cord compression were found at the time of initial diagnosis. Spinal cord compression occurs much less often in Hodgkin's disease [10-12]. To our knowledge, the case described here is only the ninth case and the third report of a patient with Hodgkin's disease who presented with spinal cord compression due to epidural and spinal disease without lymphoma

elsewhere [13-15]. Spinal cord compression due to Hodgkin's disease is very rare and usually develops in the setting of a diffuse, undiagnosed disease or at some time after the initial diagnosis.

We encountered some difficulties in determining the diagnosis of Hodgkin's disease in our case. Firstly, there was a high level of inflammatory reaction, fever, and spondylotic features in radiologic and histological findings. Secondly, histological evaluation of the spine lesion did not show RS cells, which are essential characteristics for the diagnosis of Hodgkin's disease. Finally, the inflammatory reaction and fever were temporarily improved by our antibiotic therapy, which possibly resulted from the effect of the drugs on the inflammatory cytokines produced by the tumor. In our case, the vertebral disc was maintained to the end, which we believe to be a feature of Hodgkin's disease of the spine. Thus, Hodgkin's disease should be considered a possible cause of any spondylosis-like lesion without vertebral disc invasion.

This patient represents an unusual case of primary extranodal Hodgkin's disease of the thoracic vertebrae with spinal cord compression. We successfully treated this patient with

focal field radiation therapy and systemic chemotherapy (ABVD). No further evidence of the disease has been noted elsewhere in the patient during 10 years of follow-up.

CONCLUSION

We reported an unusual case of primary, isolated Hodgkin's disease of the thoracic vertebrae with spinal cord compression. Focal field radiation therapy and systemic chemotherapy (ABVD) were effective, and showed a good prognosis. The presence of a tumor rather than inflammation should be suspected in cases when MRI confirms that the vertebral disc is maintained.

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Figure legends

Fig. 1: MRI results demonstrated that the region from the 1st –3rd thoracic vertebral body had been replaced by an osteolytic mass (left). Enhancement around the lesion and spinal cord compression were present (right). However, the lesion did not invade the intervertebral disc.

Fig. 2: Computed tomography at 6 months after admission (right) showed a larger lesion at the anterior mediastinum compared with that at the time of admission (left).

Fig. 3: Histological examination (hematoxylin-eosin [H.E.] ×10) of the biopsy specimen showed inflammatory infiltrates with fibrosis and necrosis.

Fig. 4: High-power field (×200) images showing RS cells, and lymphoma cells that were immunoreactive for CD15 and CD30.

Fig. 5. MRI at the 6 years after surgery showed no evidence of recurrence.

Fig. 6. Plain radiographs showed no abnormality 10 years after surgery.
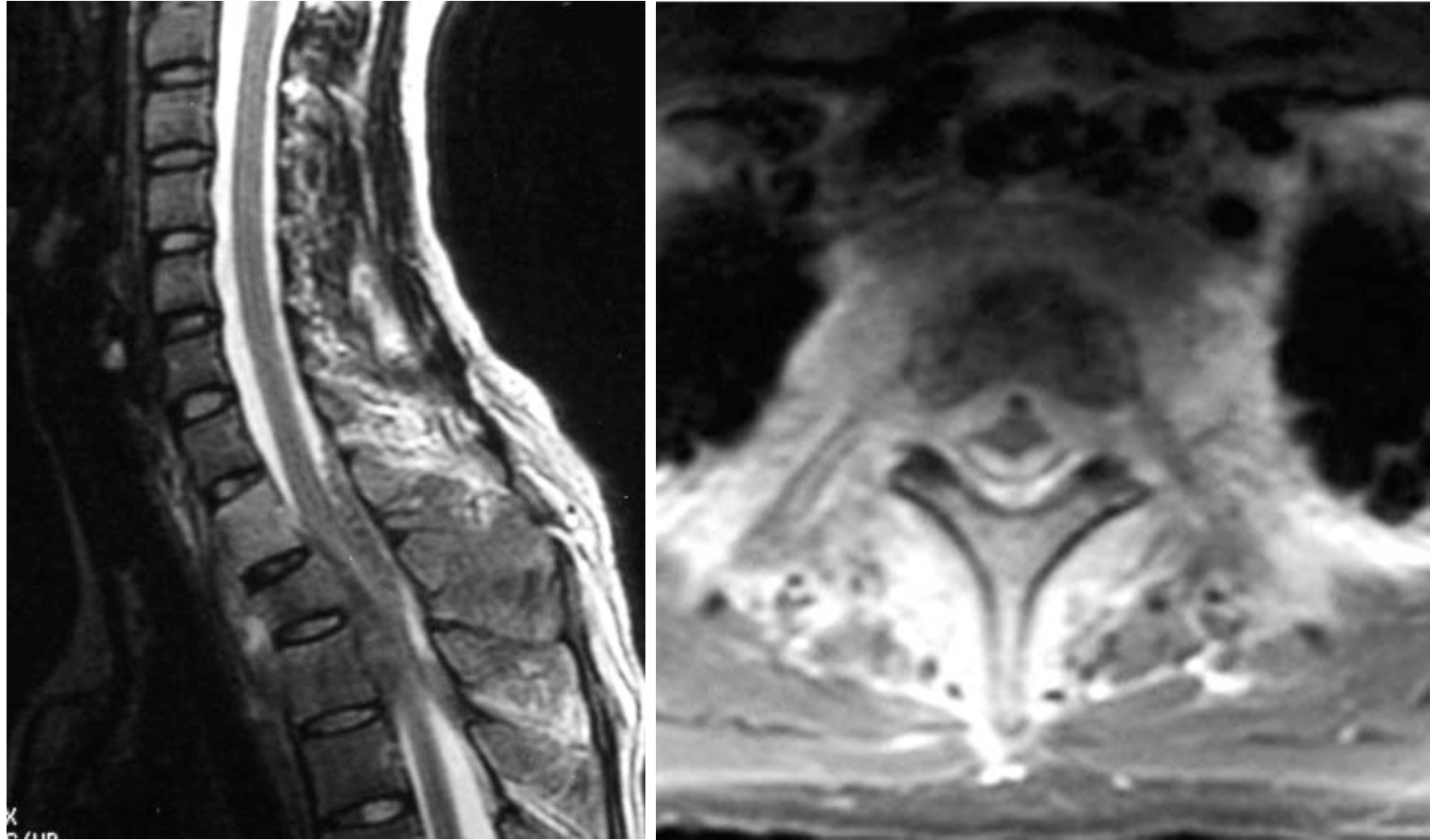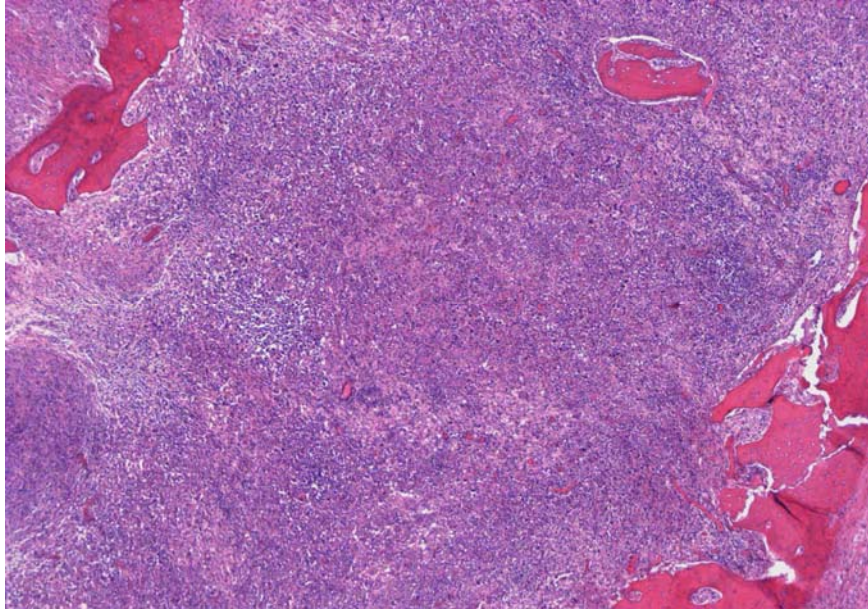
Fig. 1
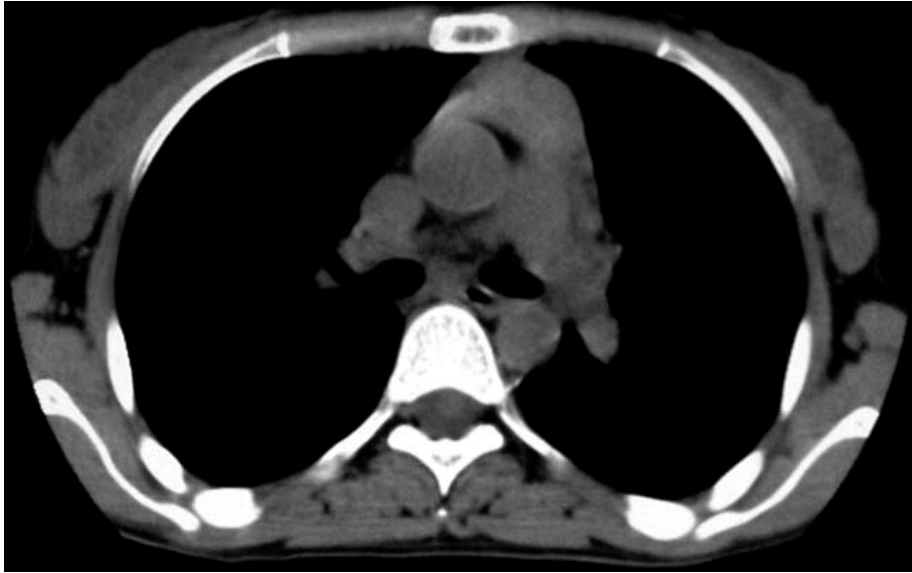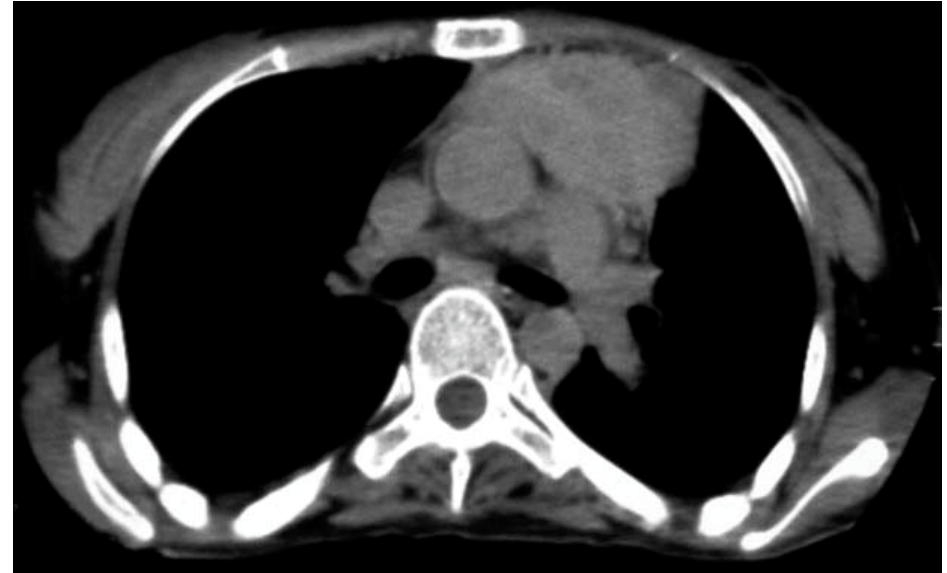
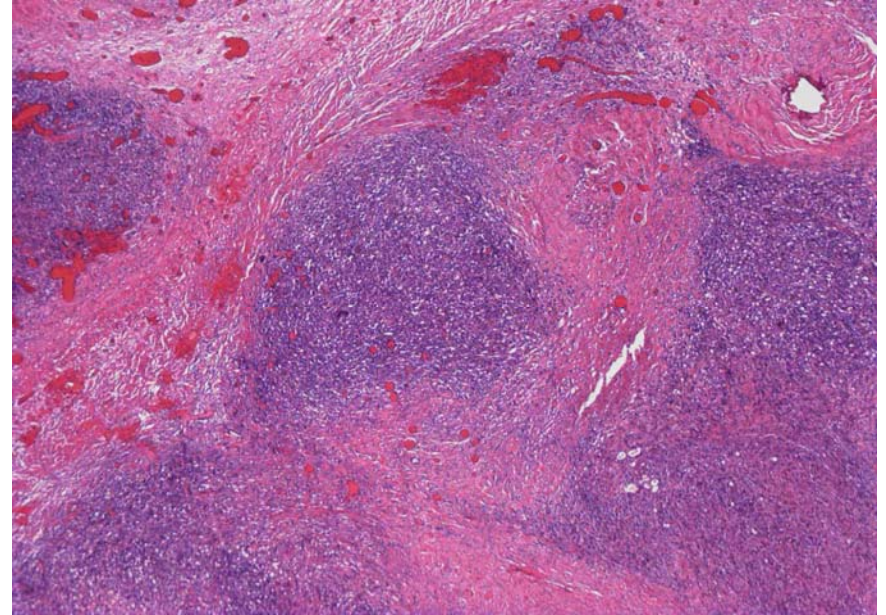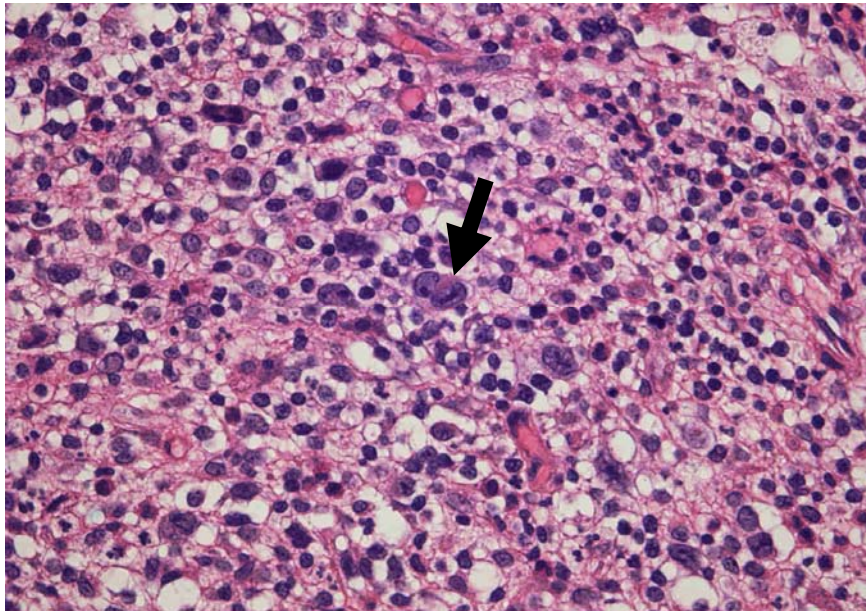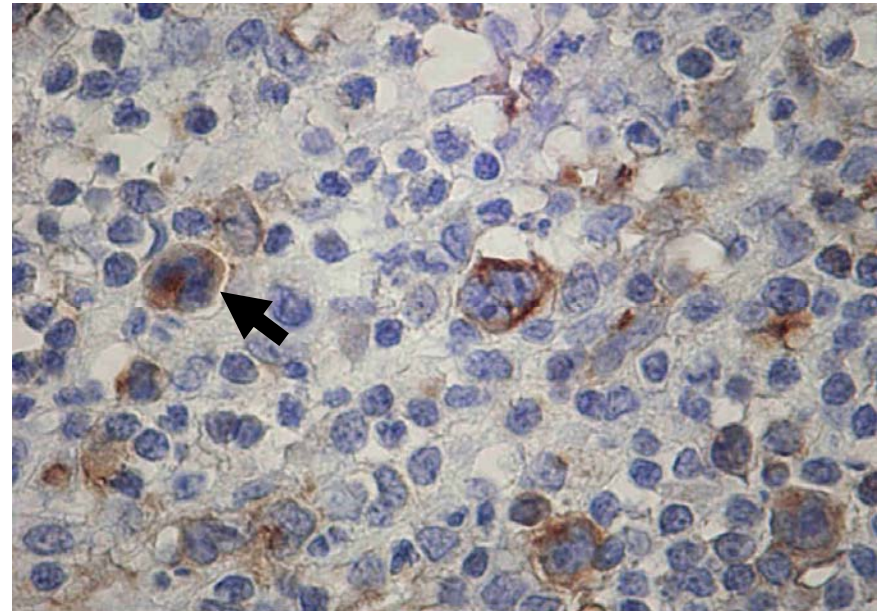On admission   6 months after admission

Fig. 2

H.E. x10   H.E. x10

Fig. 3

H.E. x100

CD30 x200

Fig. 4

Fig. 5

Fig. 6