Solitary Extramedullary Plasmacytoma Mimicking Acute Subdural Hematoma

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Key words
- Acute subdural hematoma
- Differential diagnosis
- Radiotherapy
- Solitary extramedullary plasmacytoma

Abbreviations and Acronyms
CT: Computed tomography
MM: Multiple myeloma
MRI: Magnetic resonance imaging
SEP: Solitary extramedullary plasmacytoma

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BACKGROUND: **Solitary extramedullary plasmacytoma (SEP) is a plasma cell neoplasm located outside the bone. It is rarely observed in the intracranial area. It is very difficult to diagnose this condition radiologically before surgery. In addition, dural SEP is usually misdiagnosed.**

CASE DESCRIPTION: **We report a case of plasmacytoma that presented as altered mental status after head trauma, located in the subdural area. We also describe its differential diagnosis and treatment by total removal and adjuvant radiotherapy. The 66-month follow-up findings showed distant plasmacytoma development, which was treated with radiotherapy alone.**

CONCLUSIONS: **Some pathological entities should be considered in the differential diagnosis of acute subdural hematomas. Furthermore, to the best of our knowledge, we report the first case of dural SEP mimicking acute subdural hematoma.**

INTRODUCTION

Plasma cell neoplasms are a group of disorders characterized by monoclonal proliferation of plasma cells, which secrete monoclonal immunoglobulin. The various types of plasma cell neoplasms include multiple myeloma (MM), solitary bone plasmacytoma, and solitary extramedullary plasmacytoma (SEP).[1,2] MM is characterized by multiple lesions. In contrast, solitary plasmacytoma will present with a single lesion. Solitary plasmacytoma can be divided into 2 groups according to the location: solitary bone plasmacytoma, which occurs only in the bone, and SEP, which is found in the soft tissues.[1,2] SEP represents 3% of all plasma cell neoplasms[3] and is mainly located in the upper aerodigestive tract.[4] Dural plasmacytoma is an extremely rare and unusual type of SEP.

The diagnosis of dural SEP radiologically is extremely difficult. Thus, SEP has usually been misdiagnosed as meningioma.[5-7] We report a case of dural SEP in a woman who presented with altered mental status after trauma that mimicked acute subdural hematoma.

CASE REPORT

A 48-year-old woman was admitted to our hospital's emergency department with severe headache, vomiting, and loss of consciousness after a moderate head trauma that had occurred 2 days earlier. Neurological examination revealed 4 of 5 left-sided hemiparesis with a Glasgow coma scale score of 13. The cranial computed tomography (CT) scan revealed a crescentic-shaped, homogeneous, hyperdense extra-axial mass in the right frontoparietal region extending over the cranial sutures, causing a midline shift (6 mm) with a maximum width of 13 mm (Figure 1A and B). Thus, acute subdural hematoma was preliminarily diagnosed.

Owing to the patient's altered neurological status, an emergency right frontoparietal craniotomy was performed. A gray, extra-axial, solid, hypovascular mass was detected in the subdural area. The lesion did not infiltrate into the cerebral tissue or dura mater; however, the adjacent cerebral tissue was swollen. The lesion and adjacent dura mater were totally excised. Duraplasty was performed with a galeal flap, and the bone flap was replaced. The patient's neurological deterioration gradually resolved after surgery. She experienced no complications during her recovery, and she was discharged on postoperative day 4 (Figure 1C).

Pathological examination revealed infiltration of nodular and diffusely distributed cells with slightly coarse nuclei having relatively small cytoplasm (Figure 2A). Immunohistochemistry showed that the cells were $CD38^+$, $CD3^-$, $CD20^-$, $CD56^-$, $CD79^+$, $CD5^-$, and $CD23^-$ (Figure 2B). Positive staining in favor of kappa light chain and negative staining in favor of lambda light chain were observed (Figure 2C and D). Thus, the tumor was identified as a plasma cell neoplasm.

Clinical tests were conducted for the differential diagnosis of SEP or MM. The complete blood count revealed no anemia. In addition, biochemistry revealed normal levels of serum calcium, albumin, and creatinine. No hypercalcemia or renal insufficiency was noted. The levels of β_2-microglobulin, C-reactive protein, and serum-free light chain were normal. The findings from serum protein electrophoresis and 24-hour urine collection for electrophoresis revealed no monoclonal protein. Bone marrow aspiration biopsy revealed normal histological findings without monoclonal plasma cells. A metastatic bone survey for the spine, long bones, and pelvis revealed normal results without

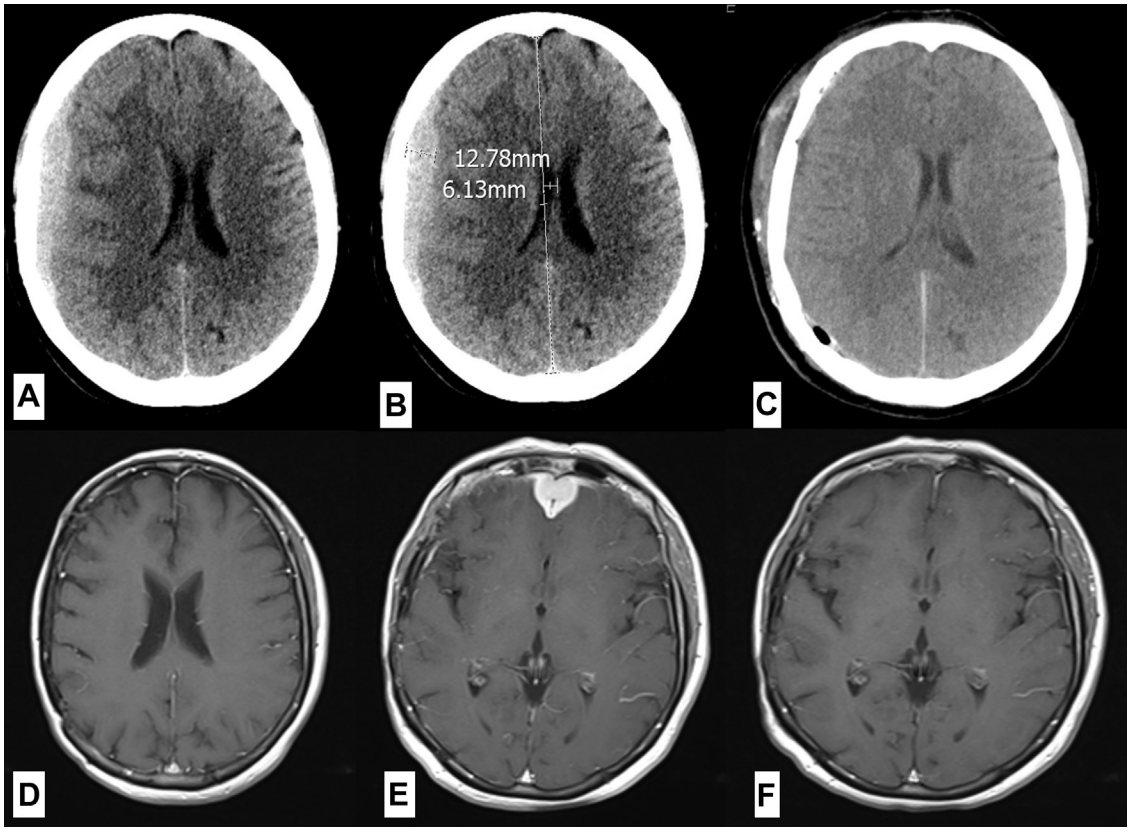

Figure 1. Radiological examination of the patient during the treatment course. (**A**) Computed tomography scan showing a subdural hyperintense lesion. (**B**) Measurements of the lesion width and midline shift. (**C**) Postoperative computed tomography scan on postoperative day 4. (**D**) Postoperative magnetic resonance imaging (MRI) study showing total removal of the lesion. (**E**) Follow-up MRI study showing distant development at 3.4 years postoperatively. (**F**) Follow-up MRI study 3 months after repeat radiotherapy to additional lesion.

any lytic lesions. Thus, the lesion was confirmed to be located in the subdural area only, and the diagnosis was confirmed as dural SEP. Postoperative magnetic resonance imaging (MRI) performed at 1 month after surgery revealed no residual lesion (Figure 1D). Six weeks after surgery, adjuvant radiotherapy was performed in 20 fractions to a total of 36 Gy to the surgical area.

The patient was followed up every 3 months for the first 2 years and every 6 months thereafter. Serum and protein electrophoresis, complete blood count tests, and serum creatinine and serum calcium tests were performed at every follow-up visit. Furthermore, cranial MRI was also performed at the original tumor site at each year of follow-up. After 3.4 years, a small (19 × 18 mm) lesion was detected in the anterior olfactory groove area. No mass effect or edema was recorded. Considering the patient's previous pathological findings and radiological features, this lesion was accepted as distant development of the dural SEP and treated with radiotherapy (35 Gy in 10 fractions in the tumor area) alone. After 3 months of radiotherapy, the lesion had disappeared as revealed by the follow-up cranial MRI study (Figure 1E). At the follow-up examination at 5.6 years, no MM progression or lesion recurrence was noted (Figure 1F).

DISCUSSION

As a subtype of plasmacytoma, SEP must be distinguished from MM for effective diagnosis.[1,2,4] Bone marrow aspirate or biopsy must be performed, and no clonal plasma cells should be observed. Moreover, no anemia, hypercalcemia, and/or renal failure should be detected from the blood test results. A metastatic bone survey or positron emission tomography/CT must be performed to confirm the absence of lytic lesions in the spine, pelvis, and long bones.[1] In the present case, this survey was undertaken, and SEP was diagnosed.

Complete surgical resection plus adjuvant radiotherapy is the definitive treatment of intracranial plasmacytomas that produce a mass effect.[8] However, small lesions diagnosed as SEP can be treated using surgery alone, without adjuvant radiotherapy if local residual tumor is not suspected.[3] In the present case, because of the patient's altered mental status, emergency tumor evacuation and removal were performed. Although total tumor removal had been achieved in our patient, owing to the large tumor diameter, adjuvant radiotherapy was also performed. SEPs are highly radiosensitive and can be treated using biopsy and radiotherapy.[9] In

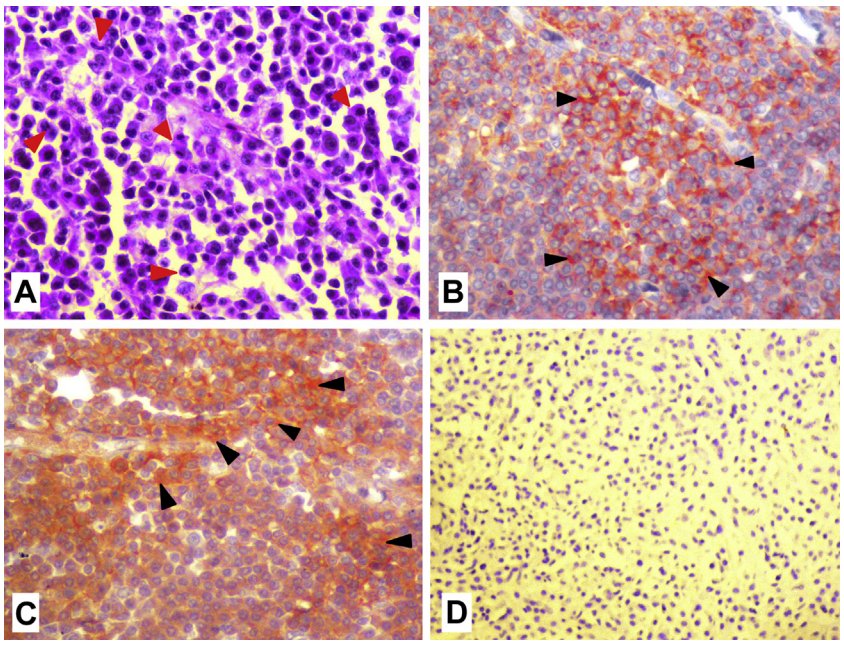

Figure 2. Microscopic views of pathologic specimens. (**A**) Hematoxylin and eosin staining (original magnification ×200). *Arrows* indicate pleomorphic plasma cells in the mitotic state. (**B**) CD38 staining (original magnification ×100). *Arrows* indicate immunohistochemical appearance of $CD38^{+}$-stained plasmocytes. (**C**) Kappa light chain staining (original magnification ×100). *Arrows* indicate staining in favor of the kappa light chain. (**D**) Lambda light chain staining (original magnification ×100) showing negative staining for lambda light chain.

a series of 18 patients with SEP of the head and neck region who underwent high-dose radiotherapy (50.4 Gy), no recurrence was noted in the radiotherapy field. One patient developed a marginal recurrence, 2 developed MM, and 4 experienced distant plasmacytoma development.[9] In our patient, a distant plasmacytoma, which was relatively small and had no mass effect, developed and was treated using radiotherapy alone.

Radiologically, the preoperative diagnosis of SEP is extremely difficult. In addition, dural SEP is usually misdiagnosed as meningioma.[5-7] In a previous case report, plasmacytoma presented as a chronic subdural hematoma.[10] CT revealed an iso-hypodense subdural mass in the frontoparietal area. The patient initially underwent surgery with 2 burr holes. However, owing to the patient's rapid deterioration after surgery, a small frontal craniotomy was performed, and a gray, hypovascular, extra-axial mass was found. Accordingly, subtotal resection and adjuvant radiotherapy were performed. Furthermore, in 2 previous reports, plasmacytoma presented as an epidural hematoma.[11,12] One of these patients presented with a biconvex hyperdense extradural mass in the frontotemporal region on CT. In addition, the lesion eroded the adjacent calvarial bone. Emergency evacuation was performed owing to the altered mental status of the patient.[11] The other patient presented with a frontal hyperdense lesion on CT after trauma. The patient underwent emergency craniotomy owing to her altered mental status.[12] A previous study investigated plasmacytoma that presented with intracerebral hematoma.[13] In that patient, L5 plasmacytoma had been diagnosed and cured 2 years previously.[13]

An intracranial plasma cell neoplasm is often accompanied by MM or develops into MM. SEPs are extremely rare in the intracranial region. In a series of 14 patients with SEP located in the intracranial region, only 1 had SEP arising from the posterior fossa dura mater.[14] SEPs rarely convert to MM. In a series of 28 cases with SEP, none converted to MM.[15] In our patient, no MM progression was noted; however, distant plasmacytoma developed and was treated using radiotherapy alone.

CONCLUSIONS

The patient's presenting history and radiological evaluations could be demonstrative for an acute subdural hematoma requiring emergency craniotomy. However, some pathological entities should be considered in the differential diagnosis of acute subdural hematomas. To the best of our knowledge, we have reported the first case of dural SEP mimicking acute subdural hematoma.

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