Solitary Intracranial Plasmacytoma Located in the Spheno-clival Region Mimicking Chordoma: a Case Report

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Solitary intracranial plasmacytoma (SIP) is very rare. This case report presents serial findings of SIP located in the spheno-clival region in a 54-year old female who presented with an inferior hemianopia in the right eye and an enlarged physiological blind spot in both eyes. Based on the initial diagnosis of a spheno-clival region chordoma, the tumour was partially resected by the nasal–sphenoidal sinus approach. Subsequently, the correct diagnosis of SIP was made based on the pathology and immunohistochemical staining of the tumour. The patient was treated using a whole skull-base radiation therapy protocol with 45 Gy and she was in good physical condition during the subsequent 22 months. The findings of a series of similar case reports documenting SIP in 20 cases published from 1976 to 2008 are also reviewed. Based on these case reports, the key features of SIP, including their clinical manifestations, clinical imaging characteristics, treatment and prognosis, are described.

KEY WORDS: INTRACRANIAL TUMOUR; SOLITARY INTRACRANIAL PLASMACYTOMA; CHORDOMA; MAGNETIC RESONANCE IMAGING; PATHOLOGY; IMMUNOHISTOCHEMISTRY

Introduction

Solitary intracranial plasmacytoma (SIP) is extremely rare.1 Only a few case reports documenting SIP have been published. The clinical symptoms and imaging features of SIP are not obvious and neurosurgeons know little about this tumour. As a result, clinicians often hesitate to reach an initial diagnosis and, therefore, to select the appropriate adjuvant therapy. A recent case of SIP is reported here and a brief review of 15 case reports documenting SIP in 20 cases published between 1976 and 2008 is also provided.1 – 15

Case report

A 54-year old female, who provided written informed consent for publication of this case, was admitted to the Department of Neurosurgery, Chang Zheng Hospital, Second Military Medical University, Shanghai, China, because of deterioration of the sight in her right eye and a mild headache that had been present for 40 days. On closer examination she was found to have an inferior hemianopia in the right eye and an enlarged physiological blind spot in both eyes. The patient’s endocrine examination was normal.
Magnetic resonance imaging (MRI) scan of the patient’s head, revealed a tumour in the spheno-clival region that showed osteolytic changes without erosion of the soft tissue (Fig. 1A – 1C). Based on the initial diagnosis of a sphenoid region chordoma, a partial resection of the tumour was performed on the patient on 20 September 2008 using the nasal–sphenoidal sinus approach. The clivus was found to have been destroyed and microscope examination revealed the sphenoidal sinus cavity to be full of tumour tissue. The tumour tissue, mixed with calcification and bone flakes, was firm and purple with an abundant blood supply. A small part of the...
tumour was resected for routine immunohistochemical examination. Residual tumour was located in the temporal base and cavernous segment of the internal carotid artery. After the operation, the sellar floor was reinforced with fibrin glue filling the sphenoidal sinus. The patient made a good post-operative recovery from the right-eye hemianopia without any surgical complications.

Histological staining with haematoxylin and eosin (Fig. 2) and immunohistochemical staining confirmed that the tumour was a plasmacytoma. Immunohistochemical staining was positive for CD138 (Fig. 3), CD38 and CD56, indicating that this tumour was a plasmacytoma. The tumour was negative for CD20, which ruled out the presence of B-cell lymphoma and it was negative for astrocytic glial fibrillary acidic protein, which ruled out the presence of glioma. Immunostaining for chromogranin A and synaptophysin were both negative, ruling out the possibility of pituitary involvement; and the staining index for Ki67 was < 1% showing that there was no active proliferation of the tumour cells.

Electrophoresis of serum for the detection of monoclonal protein and 24-h urinary light chain testing, both assays for multiple myeloma, were normal. An emission computed tomography whole body bone scan proved normal. The proportion of plasma cells in a bone marrow puncture was 1.5%, which was within the normal range. Based on these data, multiple myeloma was excluded and the diagnosis of SIP in the spheno-clival region was finally confirmed. A whole skull-base radiation therapy protocol with 45 Gy was used to treat the patient.

The patient’s visual acuity and visual field did not deteriorate further and she was in good physical condition during the 22 months following her operation. The tumour was shown not to be enlarged on a follow-up MRI scan. The proportion of plasma cells in bone marrow biopsy during follow up was

FIGURE 2: Cytological features of the single intracranial plasmacytoma showed variation from mature to immature plasma cells (haematoxylin and eosin stain)
always < 5%, consistent with UK Myeloma Forum recommended diagnostic criteria for plasmacytoma; biopsy showed 3% plasma cells in the third month and 1.5% plasma cells in the sixth month of follow-up.

Discussion

The occurrence of SIP is relatively rare, so the clinical and imaging features need to be characterized by collecting as much evidence from individual case reports as possible in order that neurosurgeons can more accurately diagnose and treat such cases. Alongside the present case report, a retrospective review of 15 case reports published from 1976 to 2008, involving 20 patients with SIP was carried out (Table 1). The key features of SIP that they report, including clinical manifestations, imaging characteristics, treatment and prognosis are described.

In terms of the epidemiological data and clinical manifestations of the 20 patients with SIP, their mean age was 57.0 years (range 18 – 82 years). According to the data shown in Table 1, the sex ratio of the patients with SIP was 1:1 (10 males, 10 females), which is different from the 2:1 ratio seen with solitary plasmacytoma of bone (SPB) reported by Soutar et al. Most of the SIPs originated in the supratentorial calvaria, especially in the frontal bone. In the skull base, SIPs were preferentially located in the orbital tip, sphenoidalis sinus, dorsum sellae-slope and clivus. At diagnosis, peripheral extramedullary myeloma is often found in the axis bones, such as the vertebral column and sternum.

The common symptoms of SIP were headache, vomiting, seizure and nerve dysfunction. Unilateral vision, visual field defects, diplopia, bone destruction, or a subscalp mass and a substantive mass in the paranasal sinuses were the specific manifestations of SIP.

The imaging features of SIP were atypical. The MRI imaging characteristics of SIP included iso- or hyperintense on T₁-weighted images, isointense on T₂-weighted images,
### TABLE 1: Summary of the data and relevant information on 20 cases of single intracranial plasmacytoma extracted from 15 case reports published between 1976 and 2008

| Case | Author | Year | Age (years) | Sex | First main symptom | Diagnosis | Location | MRI | T1/T2 | Enhanced | Arachnoid | Computed | Therapy | Follow-up | Visit |
|------|--------|------|-------------|-----|--------------------|-----------|----------|-----|-------|----------|----------| tomography | scan | visit   |
| 1    | Cerase et al. | 2008 | 67 | F | Skull destruction, Headache | Unknown | SP | Frontal bone | Orbital | ↑/– | Yes | Exist | NDA | Total removal | Stable |
| 2    | Cerase et al. | 2008 | 79 | M | Headache | Unknown | SP | Frontal bone | Orbital | ↑/– | Yes | Exist | NDA | Surgical resection | Stable for 12 months |
| 3    | Mitsos et al. | 2004 | 62 | M | Headache, vomiting | CSH | SP | Frontal bone | NDA | NDA | Exist | Skull damaged | NDA | Surgical resection | Favourable prognosis Unknown |
| 4    | Gallina et al. | 2004 | 64 | F | Headache | Meningioma | SP-κ | Frontal bone | NDA | NDA | Exist | Skull damaged | NDA | Surgical resection | Stable for 4 years |
| 5    | Gallina et al. | 2004 | 59 | F | Headache | Meningioma | SP-λ | Frontal bone | NDA | NDA | Exist | Skull damaged | NDA | Surgical resection | Stable for 4 years |
| 6    | Brannan et al. | 2003 | 61 | F | Visual loss, headache | Retinobular neuritis | SP IgG-κ | Orbital | ↑/– | Yes | Exist | NDA | Partial removal | Stable |
| 7    | Cakir et al. | 2003 | 49 | M | Orbital pain | Frontal bump | SP | Paranasal sinus | ↑/– | Yes | Exist | NDA | Partial removal | No regression in 2 years |
| 8    | Vaicys et al. | 1999 | 59 | F | Seizure | Meningioma | SP-γ | Dural | ↑/– | Yes | Exist | NDA | Partial removal | Good for 6 months |
| 9    | Vujovic et al. | 1998 | 56 | M | Seizure | Meningioma | SP-λ | Dural | NDA | NDA | Exist | Calcification | NDA | Partial removal | Stable for 4 years |
| 10   | Tanaka et al. | 1998 | 55 | M | Painless lump, Diplopia | Scalp mass | SP | Frontal bone | NDA | NDA | Exist | Skull damaged | NDA | Total removal | Stable at 7 months |
| 11   | Bourne and McLaren | 1998 | 45 | M | Diplopia | Gradening's syndrome | SP | Frontal bone | NDA | NDA | Exist | Skull damaged | NDA | Craniotomy | Stable for 8 years |
| 12   | Bindal et al. | 1995 | 51 | F | Diplopia | Pituitary adenoma | SP | Sphenoid sinus | NDA | NDA | Exist | NDA | Craniotomy | Stable for 8 years |
### TABLE 1 (continued):
Summary of the data and relevant information on 20 cases of single intracranial plasmacytoma extracted from 15 case reports published between 1976 and 2008

<table>
<thead>
<tr>
<th>Case</th>
<th>Author et al.</th>
<th>Year (years)</th>
<th>Age (years)</th>
<th>Sex</th>
<th>First main symptom</th>
<th>Initial</th>
<th>Final</th>
<th>Location</th>
<th>MRI T1</th>
<th>MRI T2</th>
<th>Enhanced T1 MRI</th>
<th>Arachnoid gaps</th>
<th>Computed tomography scan</th>
<th>Therapy</th>
<th>Follow-up visit</th>
</tr>
</thead>
<tbody>
<tr>
<td>13</td>
<td>Bindal et al.</td>
<td>1995</td>
<td>43</td>
<td>F</td>
<td>Headache</td>
<td>Meningioma</td>
<td>SP</td>
<td>Clivus</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>Craniotomy</td>
<td>No recurrence</td>
</tr>
<tr>
<td>14</td>
<td>Bindal et al.</td>
<td>1995</td>
<td>47</td>
<td>M</td>
<td>Headache</td>
<td>Unknown</td>
<td>SP-γ</td>
<td>Parietal</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>Craniotomy</td>
<td>Stable for 25 years</td>
</tr>
<tr>
<td>15</td>
<td>Bindal et al.</td>
<td>1995</td>
<td>82</td>
<td>F</td>
<td>Mental disorder</td>
<td>NDA</td>
<td>SP</td>
<td>Parietal</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>Craniotomy</td>
<td>Developed MM</td>
</tr>
<tr>
<td>16</td>
<td>Benli and Inci</td>
<td>1995</td>
<td>52</td>
<td>M</td>
<td>Frontal cranial lump</td>
<td>Cranial lump</td>
<td>SP</td>
<td>Frontal bone</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>Total removal + RT</td>
<td>Stable</td>
</tr>
<tr>
<td>17</td>
<td>Miyachi et al.</td>
<td>1990</td>
<td>61</td>
<td>M</td>
<td>Anacousia, dysphagia</td>
<td>Meningioma</td>
<td>SP-IgA</td>
<td>Unknown</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>Craniotomy + RT</td>
<td>Good recovery</td>
</tr>
<tr>
<td>18</td>
<td>Goriachkina</td>
<td>1979</td>
<td>18</td>
<td>F</td>
<td>Endocrine disturbances</td>
<td>Pituitary adenoma</td>
<td>SP</td>
<td>Sella turcica</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>Total removal + RT</td>
<td>Unknown</td>
</tr>
<tr>
<td>19</td>
<td>Gad et al.</td>
<td>1978</td>
<td>52</td>
<td>M</td>
<td>NDA</td>
<td>NDA</td>
<td>SP</td>
<td>Temporal dura</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>No cranial defect</td>
<td>Autopsy</td>
<td>Stable</td>
</tr>
<tr>
<td>20</td>
<td>Mancilla-Jimenez and Tavassoli</td>
<td>1976</td>
<td>58</td>
<td>F</td>
<td>NDA</td>
<td>NDA</td>
<td>SP</td>
<td>Meninges</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>NDA</td>
<td>Total removal + RT</td>
<td>Recurrence in 5 years</td>
</tr>
</tbody>
</table>

T1/T2 MRI, T1- and T2-weighted magnetic resonance imaging scan, respectively; F, female; M, male; CSH, chronic subdural haematoma; SP, solitary plasmacytoma; Ig, immunoglobulin; MM, multiple myeloma; ↑, hyperintense; ↓, isointense; ↓↑, hypointense; RT, radiotherapy; CT, chemotherapy; NDA, no data available.
and contrast-enhancing on gadolinium-enhanced T1-weighted scans.\textsuperscript{1, 3–6, 8, 9} It was difficult to differentiate SIP from carcinoma, meningioma and metastasis in some cases.\textsuperscript{3, 6, 7} Many authors reported that the tumours often showed a dural tail sign and some tumours even directly originated from the dura mater.\textsuperscript{3, 6, 7, 10, 12} Furthermore, nearly one-third of cases were diagnosed as meningioma during the initial diagnosis.\textsuperscript{3, 6, 7, 10, 12} As a result of their common locations, SIPs often mimicked retrobulbar tumour, optic neuritis, chordoma or pituitary tumour, chronic subdural haematoma and Gradenigo’s syndrome.\textsuperscript{2, 4, 8–10, 13} In these cases, the correct diagnosis of SIP was not made until biopsy or surgical resection.

Adjuvant therapy and prognosis of SIP differ from those of multiple myeloma.\textsuperscript{10} Some authors have suggested that meticulous examination is required to detect whether multiple myeloma has occurred.\textsuperscript{1, 5, 7, 10} Special immunohistochemical studies yielded very promising results of high prognostic value.\textsuperscript{17, 18} As Table 1 shows, where data were available most SIPs showed hyperintensity on T\textsubscript{1}-weighted MRI images.\textsuperscript{1, 3–5, 9} Multiple myelomas tend to show iso- or hyperintensity on T\textsubscript{1}-weighted MRI images.\textsuperscript{1, 19, 20} Both tumour types show no differences on T\textsubscript{2}-weighted MRI images. Of course, the final diagnosis of SIP is mainly based on pathological examination.

In terms of treatment, many authors reported that they totally resected the tumour when it was in the supratentorial or brain convexity.\textsuperscript{1–3, 8, 10, 11} For most SIPs located in the skull base that were deep with complex neural and vascular structures, limited or partial resection was usually all that was possible.\textsuperscript{3–7} Close follow-up and radiotherapy are the main treatments designed to prevent the progression of SIP to multiple myeloma.\textsuperscript{2–12, 15} SIP is a highly radiosensitive tumour and 40–50 Gy fractionated radiotherapy is recommended by several authors.\textsuperscript{12, 15–17} Once multiple myeloma occurs, chemotherapy or stem cell transplantation is necessary.\textsuperscript{21, 22} High-dose chemotherapy for multiple myeloma, such as melphalan–prednisone,\textsuperscript{1} cyclophosphamide–vincristine–dorubicin–methylprednisolone (CVAMP)–dexamethasone,\textsuperscript{21} and alkylating agents,\textsuperscript{4, 17} may significantly reduce the tumour mass.

The prognosis of the 20 patients with SIP in the follow-up period showed that one patient had progressed to multiple myeloma, one patient had a recurrence of the original tumour, three patients had an unknown status and 15 patients had not experienced either a tumour recurrence or an increase in tumour volume; one of these 15 patients had been in a stable condition for 25 years. Tanaka et al.\textsuperscript{8} reported that SPBs progress to multiple myeloma 7–23 years after presentation. Although SIP is less likely to progress to multiple myeloma than SPB,\textsuperscript{16} close follow-up is necessary for all patients.

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Conflicts of interest
The authors had no conflicts of interest to declare in relation to this article.

References


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