Case Report

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Solitary Intracranial Plasmacytoma in an Unusual Location – A Case Report

Plasmacytoma is rare form of plasma cell dyscrasias, where there is malignant proliferation of plasma cells. Solitary plasmacytoma develops in isolation without systemic manifestations of multiple myeloma (MM). We report a case of cerebellopontine angle (CPA) plasmacytoma, which masqueraded as a schwannoma with multiple cranial nerve involvement and review a literature pertaining to the same pathological entity.

Keywords: cerebellopontine angle, cranial nerve palsy, multiple myeloma, plasmacytoma,

Central nervous system (CNS) involvement in multiple myeloma (MM) is rare and accounts for only 1% of cases.[8] Intracranial plasmacytoma is a solitary myeloma plasma-cell tumor that affects the skull, meninges, and brain. It is well known that plasmacytoma, a benign lesion, may progress to multiple myeloma, a malignant and often fatal neoplasm. Plasmacytoma is rare form of plasma cell dyscrasias, where there is malignant proliferation of plasma cells. Solitary plasmacytoma develops in isolation without systemic manifestations of MM.[7] We presented a case plasmacytoma in unusual location.

Case report:

A 50-year-old diabetic gentleman presented with insidious onset, gradually progressive intermittent generalized headache associated with numbness over right half of the face for a year. He noticed diminished hearing on right ear with tinnitus, double vision and difficulty in swallowing for 2 months. He however did not have deviation of face or weakness of extremities. Examination revealed diminution of visual acuity on both eyes with lateral rectus palsy on right and medial rectus palsy on left eye. Sensory diminution was present on right V1, V2 and V3 distribution with absent corneal reflex on right eye. No facial weakness was present and there was sensorineural hearing loss on right ear with absent gag reflex. There was no bony or spinal tenderness present.

MRI brain revealed homogenous extra axial mass 3.5x3.5x3.6 cm in size in right cerebellopontine angle extending into the meckel's cave, hypointense on T1W and hyperintense on T2W, not suppressing on FLAIR image with moderate heterogenous gadolinium contrast uptake (Figure. 1). Initial impression of trigeminal schwannoma was made. Intraoperative finding of 3x4x4 cm globular, firm, moderately vascular mass arising from tent and pushing the VII/VIII complex posteriorly with

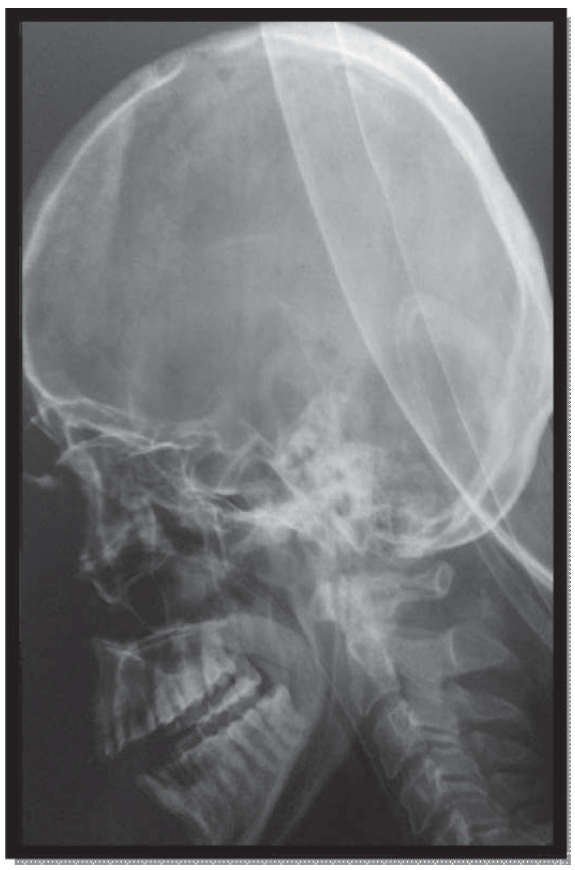

Figure 1. MRI brain revealed homogenous extra axial mass 3.5x3.5x3.6 cm in size in right cerebellopontine angle extending into the Meckel's cave, hypointense on T1W and hyperintense on T2W with moderate heterogenous gadolinium contrast uptake.

tumor capsule adherent to Vth cranial nerve was found. Histopathology revealed sheets of small round cells in uniform, monotonous population, regularly punctuated by vessels. Those cells had small round eccentric nuclei with cart-wheel distribution of chromatin and occasional paranuclear hoffs. There were no histological features consistent to schwannoma or meningioma (Figure 2). Immunohistochemical markers viz. CD45, CD138 and EMA AG were positive in tumor cells; while markers like CD 20 and CD 3 were negative in tumor cells and positive in interspersed T lymphocytes suggestive of small round cell tumor consistent with plasmacytoma.

Patient was therefore scrutinized to rule out systemic involvement and possibility of multiple myeloma association. Routine hematological examination revealed hemoglobin of 16.8 gm%, total count of 8800 with neutrophil of 55% and lymphocytes of 42%. ESR was 47 mm in 1st hour. No abnormal cells were noted in peripheral smear. Urine routine examination was normal with absent urinary Bence-Jones proteins. Blood biochemistry showed urea of 19 mg/dl, serum creatinine of 0.9 mg/

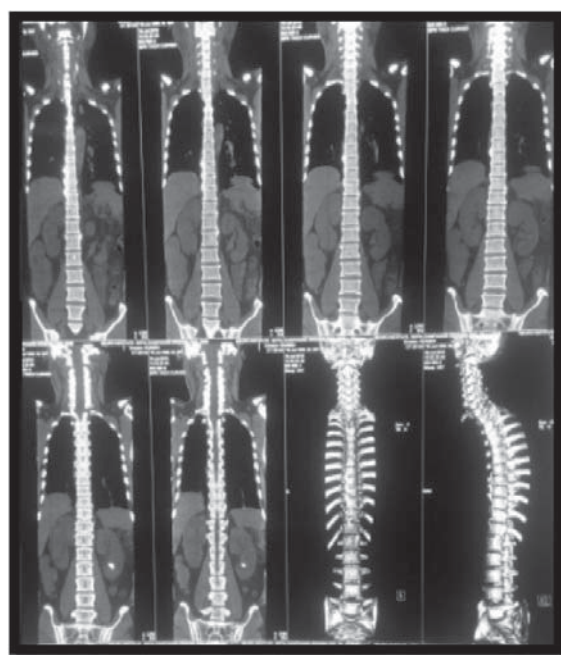

Figure 2. Histopathology: A. Medium power shows sheets of dark cells (left half) plastered against fibrocollagenous tissue of dura (right half) in X100 magnification B. High power reveals the cells to be composed exclusively of plasma cells with eccentric nuclei and paranuclear hoffs in X400 magnification. There were no histological features consistent to schwannoma or meningioma.

dl, serum calcium of 9.4 mg/dl, magnesium of 2.1 mg/dl and random blood sugar of 71 mg/dl. Cerebrospinal fluid analysis was normal without evidence of malignant cells. X-ray chest and skull were also normal CT scan whole spine was normal (Figure 3). Bone marrow biopsy was also normal. Serum electrophoresis was normal. Patient underwent gross total resection of tumor. Intra operative and post-operative period was uneventful. Postoperative, patient recovered and was subjected to radiotherapy.

Discussion:

Number of reported cases of plasmacytoma in the CNS is limited.[1] Plasmacytoma and MM represents the spectrum of the same disease, where plasmacytoma refers to the localized form and MM implies systemic dissemination. We report a case of CPA plasmacytoma, which masqueraded as a schwannoma with multiple cranial nerve involvement.

French was the first to report a solitary intracranial plasmacytoma in the hypothalamus without bone involvement or manifestation of multiple myelomatosis. This case was diagnosed as encephalitis before surgery.[3] Clarke reported a case of solitary intracranial plasmacytoma that involved the tentorium cerebelli and

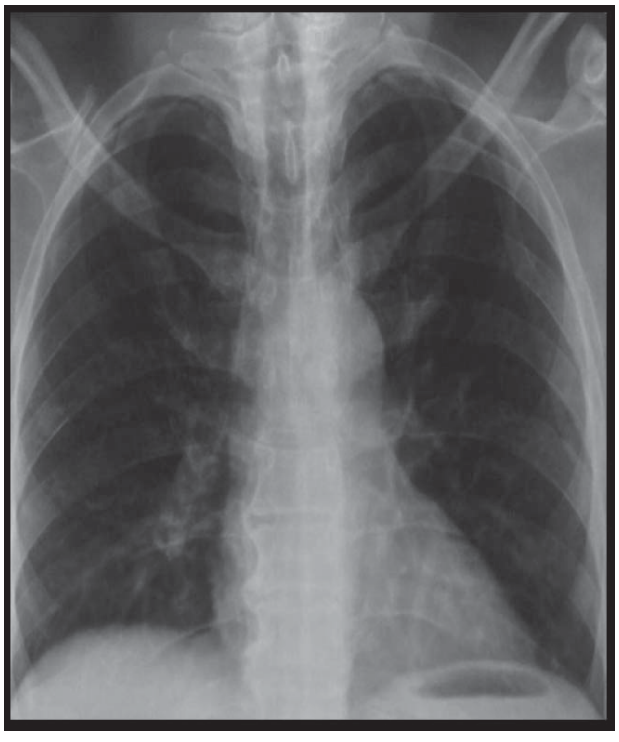

Figure 3. Chest X-ray, CT scan of Vertebra and skull X-ray didn't show any lesion suggestive of systemic pathology.

did not involve the skull.[2]Moossy and colleague reported a solitary intracranial plasmacytoma involving the frontal dura mater and falx cerebri without involvement of the cranial bone.[6] Tanaka et al. have reported a case of solitary plasmacytoma of the skull and noted that only 35 cases have been reported in literature.[9]

Silverstein and coworker studied neurologic complications in 277 myeloma patients and reported that the spinal cord compression as the most frequent (27 cases, 9.7%) presentation, while cranial nerve (CN) involvement was unusual (7 cases, 2.5%).[8] Among these seven patients one patient had isolated 6th CN palsy and another one had 3rd, 4th and 6th. Multiple CNs involvements as the initial presentation for plasmacytoma had been reported very rarely. Montalban et al. have reported an unusual case of MM with CN neuropathy due to intracranial plasmacytoma.[5] Our patient presented initially with features of multiple cranial nerves palsies and subsequent investigation confirmed the diagnosis of Cerebellopontine angle plasmacytoma without MM. Ko et al. reported a case of the intracranial plasmacytoma in the cavernous sinus presented as left 6th and partial 5th CN palsy.[4]Tappin et al. has reported a case of lower CN palsy in the form of Collet-Sicard syndrome, which turned out to be the intracranial plasmacytoma.[10]

The clinical and neuroradiological findings are generally non-specific, so they are often misdiagnosed or masqueraded preoperatively. On both CT and MRI scans, there may be mild to significant enhancement as shown in our case. Plasmacytomas involving Cerebellopontine angle are extremely rare. Since it is difficult to differentiate from CPA schwannoma preoperative suspicion of plasmacytoma should always be considered.

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