

Unusual Presentation of Pilocytic Astrocytoma

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Abstract

Pilocytic astrocytoma (PA) is the benign variant of astrocytic tumours. Pilocytic (Greek word Pilos=hair) refers to frequent presence of cells with bipolar hairlike processes. Pilocytic tumours are juvenile astrocytomas commonly presenting during first two decades of life with a lowered occurrence in adults. Cerebellum is the favoured site. Spinal cord is the rare site of presentation. We report a case of an adult intramedullary spinal pilocytic astrocytoma in a thirty four year old male.

Conclusion: An awareness of existence of spinal cord pilocytic astrocytoma is essential to facilitate an accurate diagnosis and to minimize possibility of mistaking it for a more aggressive Fibrillary or anaplastic astrocytoma. Newer modalities help in diagnosing this indolent tumour which is amenable to surgical cure.

Introduction

Pilocytic astrocytoma corresponds to term “spongioblastoma” of Zulch and Wechsler¹ and juvenile astrocytoma of Russel and Rubstein.^{1,6} It accounts for 0.6-6% intracranial tumours, 1.7-7% of glial tumours and 32% of juvenile astrocytomas.^{2,3,6} In 1931, Penfield coined the term pilocytic tumour. Depending on anatomic site and macroscopic appearance, they have a number of pseudonyms including optic nerve glioma, hypothalamic glioma, cerebellar astrocytoma, exophytic /brainstem glioma, and cystic astrocytoma.⁸ Harvey Cushing⁴ studied 76 cases and was the first to describe this entity in detail.^{2,6} These are discrete, slow growing WHO grade I tumours.⁴ They have been found at all levels of CNS axis including optic nerve, hypothalamus, cerebellum, brainstem and rarely in spinal cord.⁵ With their indolent biologic behaviour they carry highest survival.⁶ In spite of indolent behaviour they present numerous oddities. They are well

circumscribed, yet infiltrate the surrounding brain. They enhance intensely, sometimes with a ring-like pattern which is more commonly seen in highly malignant tumours. They can even produce widespread dissemination yet, they are not high grade gliomas which seem incongruous for brain tumours with a slow growth and bland histological criteria.⁶ Even more fascinating is the metastatic spread without increased mortality, commonly seen in patients with metastatic high grade gliomas. Therefore, Pilocytic tumours are an “exception to the rule”.⁶

Case Report

A 34 year old male, a manual labourer, came for tingling numbness in all four limbs and weakness in both upper limbs since two months. Local treatment did not relieve his symptoms however he could perform his daily activities with ease. There was no history of trauma or fall, fever, cranial nerve deficit or significant past history of any systemic disease.

On investigation, MRI (Fig.1) showed a moderately enhancing intramedullary SOL at C3-C7 vertebral level, perilesional oedema and a cystic necrotic area with a proximal syrinx suggesting a diagnosis of Astrocytoma. Both upper limbs showed

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Fig. 1 : T2 W sagittal image shows a heterogeneously hyperintense intramedullary lesion extending from C3 to C7 vertebral levels with a cystic component at C6 and C7 levels on the left and T1W Fat saturated post gadolinium image shows mild enhancement along the periphery of the lesion on the right.

electrophysiologic evidence of lower motor neuron type denervation in right C7-8, T1 and left C8-T1 myotomal muscles. He underwent C3-C7 laminectomy with excision of tumour.

Pathological Findings

Gross : We received multiple fragmented grey white tissue bits totally aggregating 2.5 x 2 cm along with one ml of suction material.

Microscopy: Histological picture showed a tumour comprising of fascicles of elongated cells with bipolar cell extensions, ovoid nuclei, and scanty eosinophilic cytoplasm alternating with loose microcystic areas on a fibrillary background (Fig. 2). Anaplasia and occasional mitotic figure were seen. Many Rosenthal fibres, glomeruloid proliferations and hyalinised vessels with areas of coagulative necrosis were noted. Diagnosis of low grade Pilocytic Astrocytoma of spinal cord was rendered.

Discussion

Hypothesis of a common origin from subependymal glia of all supratentorial as well as infratentorial PA is supported.¹ There is no sex predilection with peak age of presentation being 1-13 years² although they do occur in adults. Adult PA are seen in cerebral hemispheres. Spinal astrocytomas are

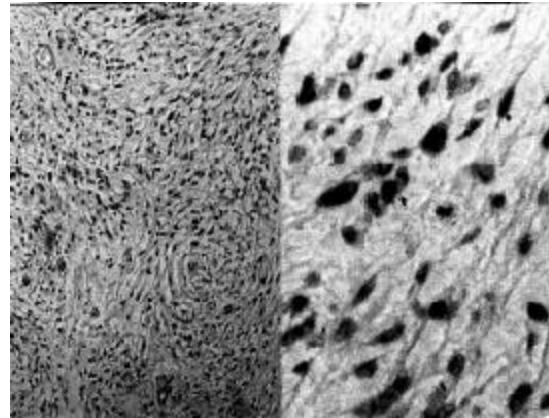


Fig.2 : Histologic picture to show biphasic pattern of pilocytic cells alternating with microcystic areas on fibrillary background on the left. High power to demonstrate bipolar extensions of pilocytes on the right.

ten times less common than cerebral astrocytomas.¹⁰ Our case presented as an adult intramedullary SOL. Clinical presentation of PA varies with site. Presenting symptoms in spinal cord include tingling numbness, backache and ataxia.^{3,10,11} Our case had similar complaints.

Morphologically, PA presents a typical constellation of gross and microscopic picture. 90% of cerebellar pilocytic tumours have a characteristic unilocular or multilocular cystic appearance, with a mural nodule.⁷ Spinal cord pilocytic tumours are intramedullary, solid and well circumscribed with distinct margins.^{3,10} Imaging studies show contrast enhancement as characteristic feature of spinal PA, which is a major factor to distinguish it from other spinal cord lesions.³ An important feature of spinal astrocytoma is tumour associated syrinx which occurs more commonly with low grade tumours.¹⁰ Our case showed similar imaging features but showed a cystic component. Histologically similar features of intracranial PA are seen in spinal cord PA^{6,8,9} showing a biphasic pattern of compact piloid tissue with dense

fascicles of bipolar cells alternating with loose glial tissue punctuated by microcysts, vacuoles and macrocysts. Calcification is an unconstant finding. Rosenthal fibres are commonly seen in dense areas and eosinophil granular bodies / protein droplets which are a constant diagnostic feature, suggest benign nature of the tumour.^{8,9} Similar histological features were noted in our case. From surgical standpoint of view, treatment of choice is radical resection which generally achieves cure.^{1,2} Gross appearance of cystic or solid tumour does not have a predictive value. Histological grade is strong prognostic indicator.^{10,11,12} Several factors are associated with its tendency to recur. Four criteria of Dumos-Duport system including nuclear atypia, mitoses, endothelial proliferation and necrosis may be present but do not imply malignancy.⁶ Such pathological findings modify prognosis and may be associated with an early recurrence.² An analysis by Lucio Palma¹ in 51 cases with a long term followup states that total extirpation of mural tumour had a better outcome.¹ Partial resection could result in recurrence.⁷ Malignant transformation is unusual. Leptomeningeal spread of low grade spinal gliomas is an uncommon event, however Abel *et al*¹¹ have reported intracranial seeding into subarachnoid space, after two years of resection, in a two year old child of spinal PA, with a six month follow-up showing stable metastatic disease.¹¹

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