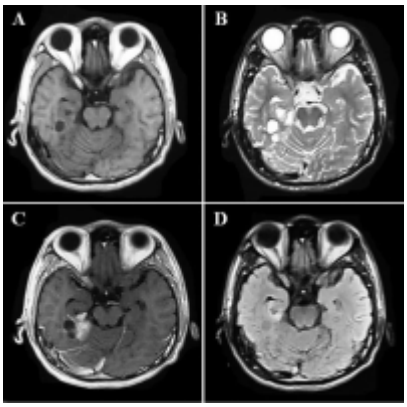
Pigmented variant of pleomorphic xanthoastrocytoma



Pleomorphic xanthoastrocytoma (PXA) is an uncommon, long-term epilepsy associated tumor of young adults. Its pigmented variant is exceedingly rare, with only six reported cases [1)].

Poyuran et al. reported the sixth case of pigmented PXA in a 24-year-old lady presenting with long-standing seizures. The MRI revealed a solid cystic lesion located in the right medial temporal lobe. Histopathologically, the superficially located tumor showed typical features of PXA with melanin-laden astrocytic component and Absence of BRAF p.V600E mutation [2)].

Gupta et al. reporteds a unique case of pleomorphic xanthoastrocytoma (PXA) in a 19-year-old male presenting with the chief complaint of seizures. On radiology, the tumor was located in the temporal lobe. It was cortically based and solid cystic in nature. Light microscopy showed pleomorphic large polygonal cells with inclusions, nuclear clustering, lipidization, and foamy cytoplasm intermingled with spindle cells arranged in sweeping pattern and focally containing cytoplasmic brownish black pigment. The pigment stained black with Fontana-Masson stain and bleached with potassium permanganate. Gomori silver stain showed reticulin fibers surrounding individual tumor cells as well as groups of cells. On immunohistochemistry, tumor cells were positive for GFAP, S-100 and focally for synaptophysin and CD34 but negative for HMB-45. CD34 revealed a specific membranous pattern around individual cells as well as groups of cells along the fibers replicating a reticulin pattern. The ultrastructural examination showed supporting melanosomes, thus confirming the melanin pigment. Sequencing for BRAF V600E showed a heterozygous mutation. This case provides further insight into the origin and pathogenesis of pigmented astrocytic tumor, additionally highlighting the characteristic CD34 staining pattern [3)].

A 16-year-old male teenager presented with seizure and loss of consciousness for 20 min. Magnetic resonance imaging demonstrated a mass occupying the right medial temporal lobe. Histological examination revealed a non-pigmented area with spindle-shaped and large xanthomatous pleomorphic cells and a pigmented region with pigmented neoplastic cells with fascicular arrangement. Immunohistochemical studies showed the tumor was positive for GFAP and low index of

Ki-67. Considering the patient's history, clinical data and pathological findings, they rendered a pigmented pleomorphic xanthoastrocytoma [4].

A 15 year old male presented with hydrocephalus from a tectal mass obstructing the cerebral aqueduct and upper fourth ventricle. The solid-cystic partly enhancing mass proved to be a pigmented pleomorphic xanthoastrocytoma, the third such example reported. The lesion revealed typical features of a PXA with the unusual addition of intracytoplasmic melanin in select lesional cells. Melanin pigment production is uncommon in glial tumors and of uncertain significance. The present case is recurrence-free one year post-operatively [5].

A large suprasellar, partly cystic, contrast-enhancing tumor was resected from a 19-year-old woman who presented with bitemporal visual field defects and reduced visual acuity. Grossly, the tumor was brown and located in the subarachnoid space. Histologically, it was composed of spindle and pleomorphic cells, including giant tumor cells, with markedly pleomorphic nuclei. Reticulin fibers surrounded single cells and small groups of cells. Very few mitotic figures were found in the tumor, and no necrosis or microvascular proliferation was seen. The tumor thereby resembled a pleomorphic xanthoastrocytoma. Many of the tumor cells contained a dark-brown intracytoplasmic pigment, shown to be melanosomal melanin by ultrastructural examination. Immunohistochemical examination demonstrated that the pigment was present in glial tumor cells. Only four cases of pigmented astrocytic tumors have been published, none of these were suprasellar. The patient received fractionated radiotherapy with a total dose of 48.6 Gy 14 months after gross total removal of the tumor. She is alive without relapse after 12-year follow-up [6].

A 32-year-old man who presented with partial complex seizures. Radiologically, the mass was located in the medial temporal lobe and was solid and cystic. Microscopic examination revealed features of a pleomorphic xanthoastrocytoma with some heavily pigmented cells. The pigment was demonstrated to be melanosomal melanin, which was confirmed by special stains, immunohistochemistry, and electron microscopy [7].

References

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