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(Metastatic) Multifocal meningeal melanocytoma of anterior cranial fossa: "a unique case report "and review of literature.

KEYWORDS: Melanocytoma, anterior cranial fossa, Metastases.

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ABSTRACT *Melanocytomas are borderline tumors between Nevus and melanoma (1,2,4).Posterior fossa, base of skull and spinal cord are most frequently reported sites in the nervous system. Tumors are very rare in the anterior cranial fossa, but very few cases of meningeal Melanocytomas reported in the literature (2). Metastases is extremely rare in Melanocytomas, however pigmented epethelioid Melanocytomas (PEM) are known to produce nodal metastases(3). Ali et al(4) Poalo Merciadri(5) published two different cases of Multifocal Meningeal Melanocytomas from spine, spread to the CNS through CSF.The aim of this report is, this is the first case, to the best of our knowledge, reporting metastases in Melanocytomas. A 47 years female patient, who is a house wife presented with complaints of head ache, vomitings and weakness of right side of the body. She was evaluated in the neurosurgery department, found to have anterior cranial mass by contrast enhanced CT, and underwent total excision. Post-operative histopathology suggested melanocytoma. Post-operative period went uneventful, patient has improved symptomatically. Post-operative MRI showed multiple sites of brain metastases. We found retrospectively Nevus over anterior chest wall, which has been sent for excision biopsy, however Nevus did not shown any features of melanoma on pathological examination.Conclusion: Melanocytomas of anterior cranial fossa are very rare. Although they are benign (1, 2, 4) locally aggressive, and nodal metastases is reported from PEM in few case reports. Metastasis is extremely rare.*

Case details:

A 45 years female, who is a house Wife, reported to JIPMER with complaints of hemiparesis and raised ICT. We found large mass in the anterior cranial fossa on CT, which was compressing temporal and frontal lobe.(Figure1a,b&c). She underwent emergency, and total excision of tumor. Post-operative period went uneventful.PreOP CT: a) plain CT Showed well defined enhancing mass in anterior cranial fossa. b)CECT also Showed contrast enhancement of tumor. c) Post op CECT Showed post op edema. (Figure1a,b&c).Post op MRI: T1w Images Showed hyper intense multiple peripherally enhancing lesions with gradient hypo intensities in bilateral parietal, occipital and right temporal lobes. Suggestive of metastatic lesions with hemorrhage. Post-operative changes in right frontal lobe with focal meningeal enhancement.(Figure 2a&b).Histopathology Showed tumor arising from meninges(Fig 3). Immunohistochemistry (IHC) positive for HMB 45, negative for GFAP and Ki-67 is Very low <1%. (Fig 4).

Patient had a Nevus over the anterior axillary fold, which has been sent for histopathological examination.Sections through subcutaneous fat revealed no tumor. There is no evidence of melanocytic proliferation or tumor cells (Figure5). In view of metastatic disease, patient has considered for Palliative whole brain radiotherapy 30 Gy in 10 fractions.She completed treatment without any significant morbidity. On follow up patient had partial regression of the lesions, however we need longer follow up period to comment on response. On further up, patient presented with raised ICT and back pain.Plain CT showed residual lesion in the brain and surprisingly, we found multiple lesions in the vertebrae. (Figure 6a,b,&c).

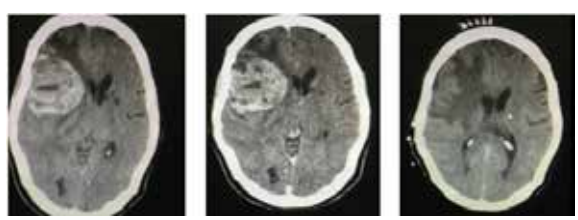

Figure 1 a, b&c PreOP CT: a) plain CT showing well defined enhancing mass in anterior cranial fossa. b) CECT also showing contrast enhancement of tumor. c) Post op CECT showing edema.

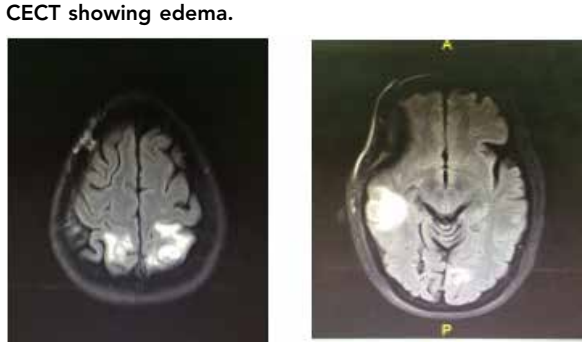

Figure 2a&b Post op MRI: T1w Images showing hyper intense multiple peripherally enhancing lesions with gradient hypo intensities in bilaterally parietal, occipital and right temporal lobes suggestive of metastatic lesions with hemorrhage.Post-operative changes in right frontal lobe with focal meningeal enhancement.

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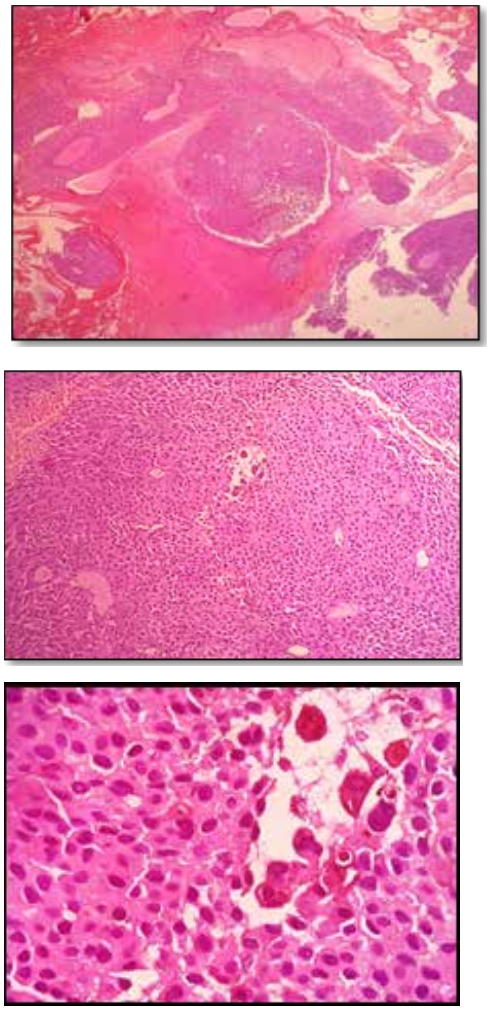

Fig 3: (a) Histopathology shows tumor arising from meninges (H&E x 100). (B) Tumor arranged in nests (H&E x 200). (C) Spindle cells with oval nuclei and few cells show cytoplasmic brown granular melanin pigment (H&E x 400).

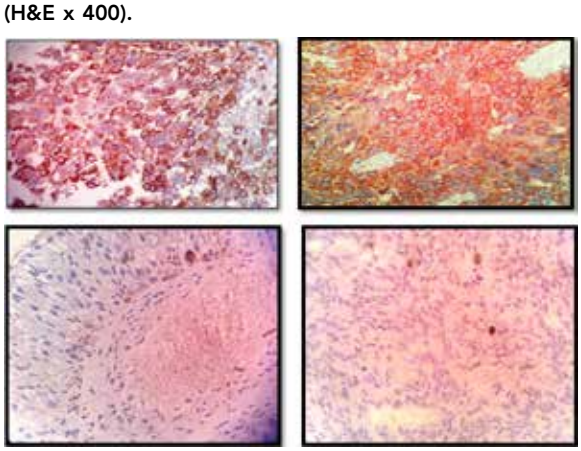

Fig 4: Immunohistochemistry (IHC) with (A)HMB 45 (X100) and (B) MelanA (X100) shows cytoplasmic positivity (C) IHC with GFAP (x100) negative in tumor cells (D) IHC with Ki-67 (x100) very low <1%.

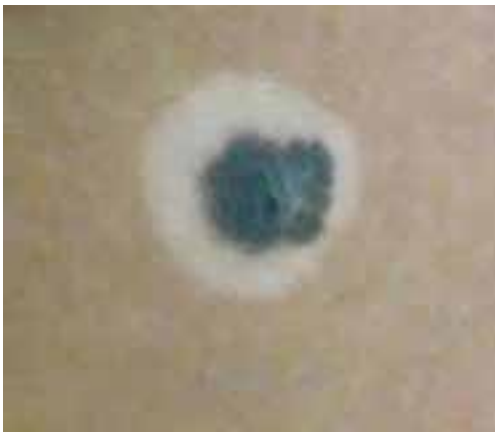

Figure5: shows nevus over anterior axillary fold.

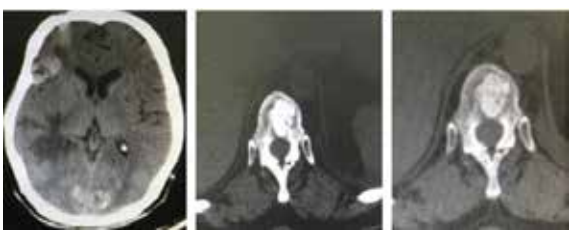

Figure 6: a,b,& c showing residual disease in the parietal and occipital and temporal lobe. Multiple metastases over D5 & L1 vertebrae.

Discussion:

Melanocytoma was first described by Limas and Toi in 1972(6), the tumors are benign usually arises from central as well as peripheral nervous system. The incidence is very very low 1 case per million people, shows its rarity (7).The diagnosis is mainly by Histological examination. The exact etiology is unknown,leptomeninges are the site of origin in central as well as peripheral neuraxis.(1,4,5).Most commonly involved sites are the base of the brain, the cerebellopontine angle, the pineal body and spinal cord (2,8).Approximately one third of the cases arising from spinal cord especially from intramedullary compartment, and rarely from extramedullary compartment(8,9).

Anterior cranial fossa Melanocytomas are very rare, very few reports were available in the literature. Kawaguch et al.(10) reported a case of meningeal melanocytoma located in the region [8]. In 2003, Uozumi et al.(11) presented a case of meningeal melanocytoma from frontal lobe in a recurrent set up. Meningeal Melanocytomas are frequently reported in the literature, Hino et al, reported a patient with a combination of nevus of Ota and meningeal melanocytoma. Piercecchi- Marti et al reported another case of meningeal melanocytoma from temporal lobe associated with Nevus Ota (13).

Symptoms depends on site of involvement, patients May presented with headache, vomiting, neuropsychiatric symptoms, spinalcord compression, and rarely seizures(4,14). our patient presented with hemiplegia with seventh cranial nerve involvement. The diagnoses of Melanocytomas are always on histopathological examination, (14, 15) because the clinical and radiological features are not definitive for Melanocytomas. Meningioma often confused with melanocytoma on radio graphically. The tumor appears hyper or iso-intense on CT scan with contrast enhancement, and presents with a high signal on T1- and fluid attenuated in-

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version recovery (FLAIR), and a low signal on T2-weighted MRI with contrast-enhancement (14,15).

Grossly, the tumor typically presents like a black lesion that is attached to the underlying meninges. Microscopically, melanin granules are commonly seen. Immunohistochemically, meningeal melanocytomas are positive for S-100 protein, HMB-45 and Vimentin (4, 5, 14, 15). Keratin, EMA and Glial Fibrillary Acidic Protein (GFAP) are usually negative; Positive HMB-45 and negative EMA are strongly indicates melanocytoma (20,21). Our case also shown similar immuno histochemistry, and low mitotic rate, hence we ruled out Melanoma in this case.

Surgery is the primary modality of treatment (4,5,14). Different approaches have been defined in the literature especially for anterior cranial fossa. Our patient underwent FT craniotomy and gross total resection. The role of adjuvant radiotherapy is still debatable after total excision of tumor.

However reports have been showed late recurrence even in completely excised tumors. There are no definitive indications for adjuvant radiotherapy, however studies were showing the importance of proliferation index like Ki-67 and MIB 1 are markers for adjuvant radiotherapy(16). Radiotherapy is a definitive option in conditions like incomplete surgeries especially base of skull, where tumor cannot be removed completely due to inaccessibility, recurrence and malignant transformation like melanoma (17, 18, 19,20). Although meningeal melanocytoma is benign, relapse and malignant transition have been reported by Wang and colleagues. Malignant transformations were also reported in spinal meningeal melanocytomas(20). In 2003, Uozumi et al (11) reported recurrence after 4 years even in gross total removal. Role of chemotherapy and targeted therapies are not well defined in the literature. Our case showed multiple secondaries in other Parts of brain parenchyma, and vertebrae. In contrast to, the previous two cases reported and in the literature, our case typically had vertebral body secondaries, without any iinvolvement of the spinal cord clinically and radiologically. Hence, we called, this case as a metastatic melanocytoma. This patient was considered for palliative radiotherapy to Whole brain as well as Vertebrae.

Conclusion:

Melanoma transformation, late recurrence are commonly reported in melanocytoma, but metastases from melanocytoma is ever reported event. Complete surgical removal of tumor is recommended, Radiotherapy is recommended in adjuvant as well as relapse setting.

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