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Primary intracranial malignant melanoma: A case with review of literature

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ABSTRACT

Introduction: Primary intracranial melanoma is an extremely rarely lesion of the brain.
A 33-year-old man, without co-morbidities, presented with headaches.
MRI of the brain showed a mass lesion in the left temporal lobe, with the characteristics signals of melanoma. The histology and immunohistochemistry confirmed the diagnosis after surgical excision of the lesion. The patient remained asymptomatic until the last visit of postoperative follow D + 1 year.

Keywords: *Intracranial lesions, Malignant melanoma, Prognosis*

Introduction

Primary intracranial melanomas are rarely reported tumors.
They account for only 1% of all the different forms of melanomas [1].
Primary malignant melanoma is indistinguishable from metastatic melanoma by radiological and pathological exams (2).
Primary CNS melanoma is an aggressive disease that evolves to other organs through metastases [3].
The diagnosis of primary intracranial malignant melanoma requires the exclusion of the possibility of secondary lesions. This by a clinical examination and an ophthalmologic and radiological evaluation [4].
As part of this work we present a case of temporal primary malignant melanoma in young male patients.

Observation

K.A, aged 33 years old, presented with an intracranial hypertension syndrome characterized by headache and vomiting.
His neurological examination was normal and his medical history were unremarkable.
Brain magnetic resonance imaging (MRI) revealed a mass in the left temporal region.
The peripheral cystic portion showed a long iso signal on T1, an iso-short signal on T2, and was slightly improved after gadolinium administration.
Pathological field is surrounded by an irregular edema (Fig. 1 C-H).
According to CT and MRI characteristics, can be suspected meningioma with intratumoral bleeding and hemorrhagic metastasis.

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During Intra-operative period a large irregular blackish mass was objectified in the left temporal region.
The mass was well defined without infiltration into the surrounding tissue allowing complete excision of the lesion. However, the peritumoral tissue was oedematous (Figure 2A and Figure 2B).

Figure 2A

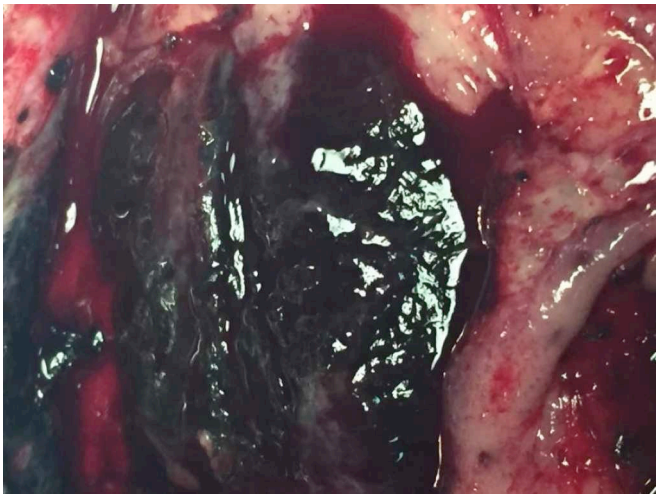

Figure 2B

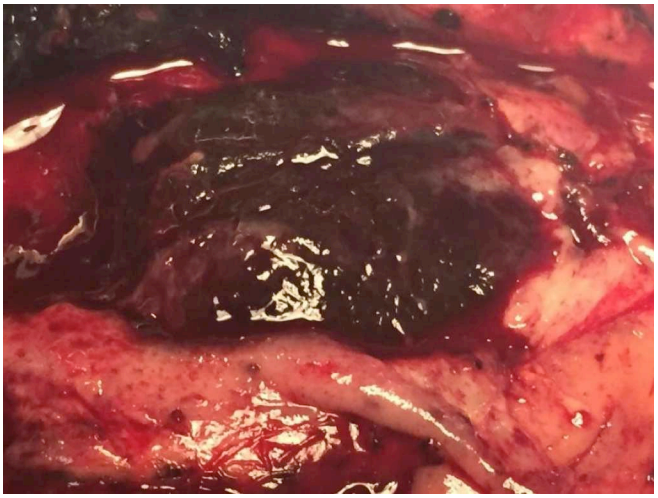

Figure 2A and Figure 2B : Images of lesion before resection

Subsequent investigations, including dermatological examination, ophthalmologic examination, endoscopy of the gastrointestinal tract, body scintigraphy, as well as whole body exploration did not reveal the presence of any sign of melanoma in other parts from the body.

Discussion

Primary cerebral melanoma is a very rare tumor whose prevalence ranges from 0.17% to 1%. Chen Y and al [1, 4].
Melanocytes are found in the skin, mucous membranes, meninges and parenchyma of the brain [1].
The sites of primary CNS melanoma by order of frequency are the hemispheres, cerebellum, medulla, cervical spinal cord and rarely at the level of the olfactory bulb. Chakraborty et al [1].
It should be noted that primitive melanomas are characterized by:
1 Single intracerebral tumors.
2 Risk of systemic metastases.
3 Development in young people under 50
Our patient had no systemic melanoma objectified by clinical examination. Thus there was a solitary mass.
According to the literature, the prevalence of primary cerebral melanoma is mainly cited higher in the following countries: Japan, Eastern United States and Europe.
Preoperative diagnosis of primary CNS melanoma is difficult except in cases where melanoma is associated with neurocutaneous melanosis or when melanin or melanin-containing cells are detected in the cerebrospinal fluid. The results of cerebral MRI of intracranial melanomas are not specific.
According to the literature, we have found 20 previously reported cases of primary intracranial malignant melanoma. **Table 1** contains the details of these cases: We documented 20 cases reported over 25-years period from 1990-2015.
A predominance is noted among male patients.
The tumor has been frequently observed in patients under 50 years old. Most cases involved the different areas of the brain, except for the three cases whose seat is the cerebellopontine angle.

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Our The symptoms present were headache, ocular symptoms, hemiparesis, and seizures.
Six patients showed recurrence mainly within 18 months, except in one case where the tumor recurred only after 23 years.
The excision of the tumor was possible among 14 cases. The other patients received radiotherapy and adjuvant chemotherapy.
Five patients died after one year. One patient survived for 23 years.
The extension of metastases was mentioned in three cases.
Age, gender, and symptoms of our case presented in this work were similar to most of the cases described in the literature.
He received chemotherapy, radiotherapy and did not show recurrence or metastases at 1 year leat.
The therapeutic consensus of primary cerebral melanomas is based on complete resections, plus effective postoperative radio and chemotherapy [7,8].
Previously, some studies have indicated that stereotactic radiosurgery (SRS) alone or conventional radiotherapy may improve the lifespan of patients with cerebral melanoma [2].

Conclusion

It is difficult to diagnose primary cerebral melanoma in the absence of any cutaneous melanosis.
A high index of clinical suspicion as well as good reports of pathology is the key to the diagnosis of these extremely rarely tumors.
The roles of adjuvant radio-chemotherapy have not been well established.
Improved targeted therapies, immunotherapies and chemotherapies could provide more effectiveness treatments for primary brain melanomas.
The case presented as part of this work is currently in remission

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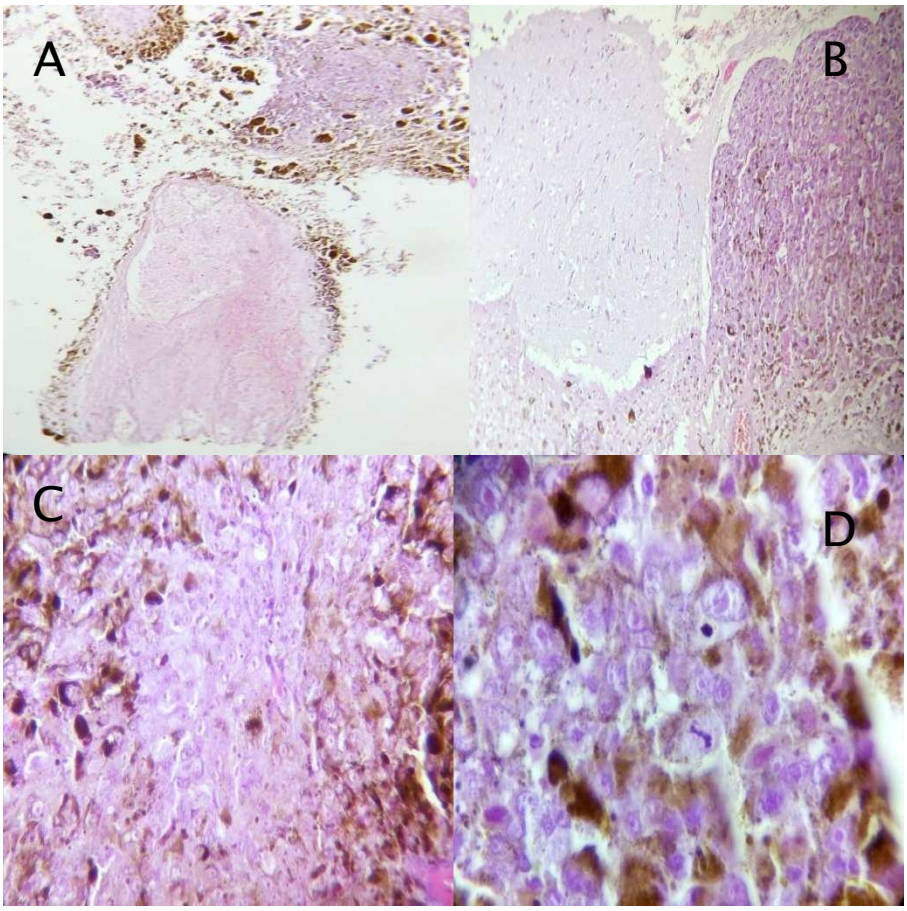

A- Cerebral parenchyma laminated by malignant tumor prolifération of hyperpigmented round cells

B- Tumor necrosis foci.

C- Massive ranges of ovoid cells with presence of intra cytoplasmic melanin pigment (arrow) (HE GX 200).

D- Prominent nucleolus characteristic (black arrow) with mitotic figures (red arrow) (staining HE GX400).

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Table 1: 20 previously reported cases of primary intracranial malignant melanoma

	Authors	Age Sex	Site	Clinical	Recur	Treatment	Survival	Other
1	Nakagawa **1989.** *al*	57 *male*	Intracranial	Persistent headache	Thrice	Excision, chemo-immuno-radiotherapy, chemo radiotherapy.	Died after 9.6 years	Nil
2	Braga et al, 1989	72 Female	CP angle	Vertigo, progressive deafness	-	Total excision	Died 5th postoperative	Nil
3	Lizuka et al, 1990	76 male	Occipital	Progressive headache, visual disturbance	+ after 8 months	Total excision	1 year follow-up alive	Nil
4	Takano et al, *1992*	34 male	Intracrania	Headache, decreased vision	-	Chemotherapy, interferon-bet.	NA	Nil
5	Lee et al. 2004.	*66 male*	Left fronto-parietal region	Headache, right hemiparesis	-	Total excision	Died after 6 months	Nil
6	2005. Ônal et al,	*38 male*	Posterior fossa mass Recur-left temporo-	Headache, vomiting, ataxia	+ after 17 years	Excision, adjuvant external beam irradiation, chemotherapy, and-interferon chemotherapy	17 years	Diffuse intracerebral mets at recurrence
7	Bhandari et al 2010	29 male	CP angle	Vertigo, headache, lower cranial nerve involvement	after 10 months	Subtotal excision with adjuvant radiotherapy	Died after 1-year	Nil
8	Azar et al *2010*	*21-male*	Parietal	Headache, mild left hemiparesia	-	Total excision, radiotherapy and chemotherapy	1-year follow-up alive	Periorbital blue nevus
9	Shah et, al 2012	28 Female	Left temporal lobe	Headache, numbness of the left side of face and diplopia	+ after 23 yeas	Excision of mass	.4 years alive	Nil
10	Mahajan et, al 2013	55 Female	Clivus	Headache, decreased vision,	-	Palliative chemotherapy and brain irradiation	NA	Skeletal mets
11	Ponni *et al.,* 2014	38 *male*	left CP angle	Deviation of left eye, headache	-	Total resection, adjuvant radiation and chemotherapy	1 vea r follow-up alive	Cerebellar infiltration
12	*Wang et, al 2014*	8 patients	Intracranial	Headache, decreased vision, vomiting	1 pt- + after 16th month	Total excision-6 Subtotal excision-2, radiotherapy-6	13.8 months follow-up: 3died, 5 alive	Nil
13	Suranagi et, al.)Present case) 2015	65 *male*	Right parafalcine frontal region	Headache, seizures, hemiparesis	-	Total excision	1- year follow-up, alive	Nil

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