Case Report
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Intraparenchymal Atypical Meningioma in Basal Ganglia Region in a Child : Case Report and Literature Review

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Intraparenchymal meningiomas without dural attachment are extremely rare, especially when they occur in basal ganglia region in child. An 8-year-old boy was admitted at our hospital, complaining of recurrent headache and vomiting for 3 months. Neurological examination showed impaired vision and mild paresis of the left extremities. Magnetic resonance imaging demonstrated a lesion located in the right basal ganglia region extending to superasellar cistern with solid, multiple cystic and necrotic components. Computed tomography revealed calcification within the mass. Due to the anterior cerebral artery involvement, a subtotal resection was achieved and postoperative radiotherapy was recommended. Histopathological examination indicated that the lesion was an atypical meningioma. The postoperative rehabilitation was uneventful. Mildly impaired vision and motor weakness of left extremities improved significantly and the patient returned to normal life after surgery. To our knowledge, intraparenchymal atypical meningioma in basal ganglia extending to superasellar cistern was never reported. The significance in differential diagnosis of lesions in basal ganglia should be emphasized.

Key Words : Intraparenchymal meningioma · Basal ganglia · Atypical meningioma.

INTRODUCTION

Meningiomas are the most common benign tumors among central nervous system (CNS) neoplasms[1]. However, meningiomas are rare in childhood and adolescence, representing only 0.4–4.6% of all CNS tumors[5]. Those located in intraparenchymaare much more rare without dural attachment[3].

Intraparenchymal meningiomas, also considered as the same type of subcortical meningiomas, are defined as meningiomas located in brain parenchyma without dural attachment, occasionallyreaching the brain surface[2,4,6]. To our best knowledge, there are only 18 intraparenchymal meningiomas in children reported in the literature, with most of them in the cerebral lobes[2,6-20].

Here we report the first case of primary intraparenchymal meningioma in the deep basal ganglia region in children and review the pertinent literature to discuss the clinical presentation and management, radiological features, and possible pathogenesis. The significance in differential diagnosis of lesions in basal ganglia is also emphasized.

CASE REPORT

An 8-year-old boy presented at our hospital complaining of

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recurrent headache and vomiting for 3 months. The patient saw a doctor in a clinic before admission to our hospital and took some medicine for cold. The symptoms got relief temporarily but became severe 1 week ago.

Neurological examination showed impaired vision and mild paresis (IV- grade of muscle strength) of the left extremities. CT (Fig. 1A, B) revealed an iso- to hyperdense lesion in the right basal ganglion extending to superasellar cistern, 4.0× 4.4 cm in sizewith peripheral calcification. No hyperostosis of sellaturcica was noticed (Fig. 1C). Magnetic resonance imaging (MRI) demonstrated most of the mass lesion was iso-intense on T1-weighted, T2-weighted and fluid-attenuated inversion recovery images with inhomogeneous enhancement (Fig. 2A-E). No duraltail sign was noted. Diffusion tensor image revealed the right pyramidal tract was partially disrupted.

Under the guide of navigation, a transcortical (through the right middle frontal gyrus) approach was adopted to explore the lesion. There was no dural attachment, but the tumor was tightly adhered with the anterior cerebral artery (ACA). Small vessels supplied the anterior aspect of the tumor, subtotal resection (STR) was achieved.

Histopathological examination revealed features of atypical meningioma with Ki-67 labeling index been approximately 10% (Fig. 3A-C). Immunohistochemistry showed the tumorpositive for epithelial membrane antigen and negative for glial fibrillary acidic protein and S-100 protein (Fig. 3D-F).

The patient suffered transient exacerbation of impaired vision and weakness of contralateral limbs after operation, which resolved significantly after 3 months follow-up. Postoperative MRI revealed that most of the tumor had been resected and the residual part has been detached from dorsumsellae (Fig. 2F).

DISCUSSION

Clinical presentation

Primary intraparenchymal meningiomas are rare, but more frequent in children and adolescents than in adults[7]. To date, only 19 patients including ours, have been reported in the English-language literatures, which are summarized in Table 1[2,6-22]. The age ranged from 0 to 18 years. Gender distribution showed a male dominance (n=14, 73.7%) in children and adolescents. The most common location was frontal lobe (n=8, 42.1%), followed by temporal lobe (n=5, 26.3%), parietal lobe (n=2, 10.5%), frontoparietal lobe (n=1, 5.3%), parietooccipital lobe (n=1, 5.3%), brainstem (n=1, 5.3%) and basal ganglia extending to superasellar cistern (our case, n=1, 5.3%). Presenting symptoms depend on tumor location and intracranial pressure, and seizure (n=13, 68.4%) was most frequent.

Management, features of histopathology and prognosis

Gross total resection (GTR) were achieved in 15 of 19 cases (78.9%). There was 70% remnant in one patient, because of the lesion in brainstem[11]. Another patient got subtotal resection, but the reason was not stated[18]. The study by Kotecha et

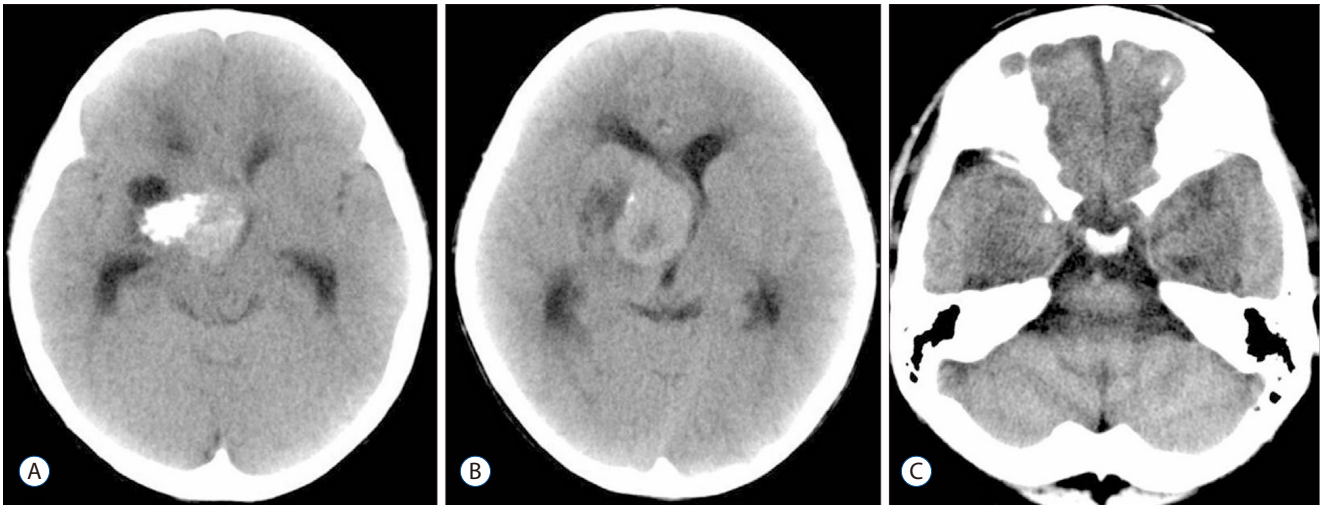

Fig. 1. CT revealing calcification, necrosis, cystic formation of the mass lesion (A and B). No hyperostosis or bone absorbtion was observed (C).

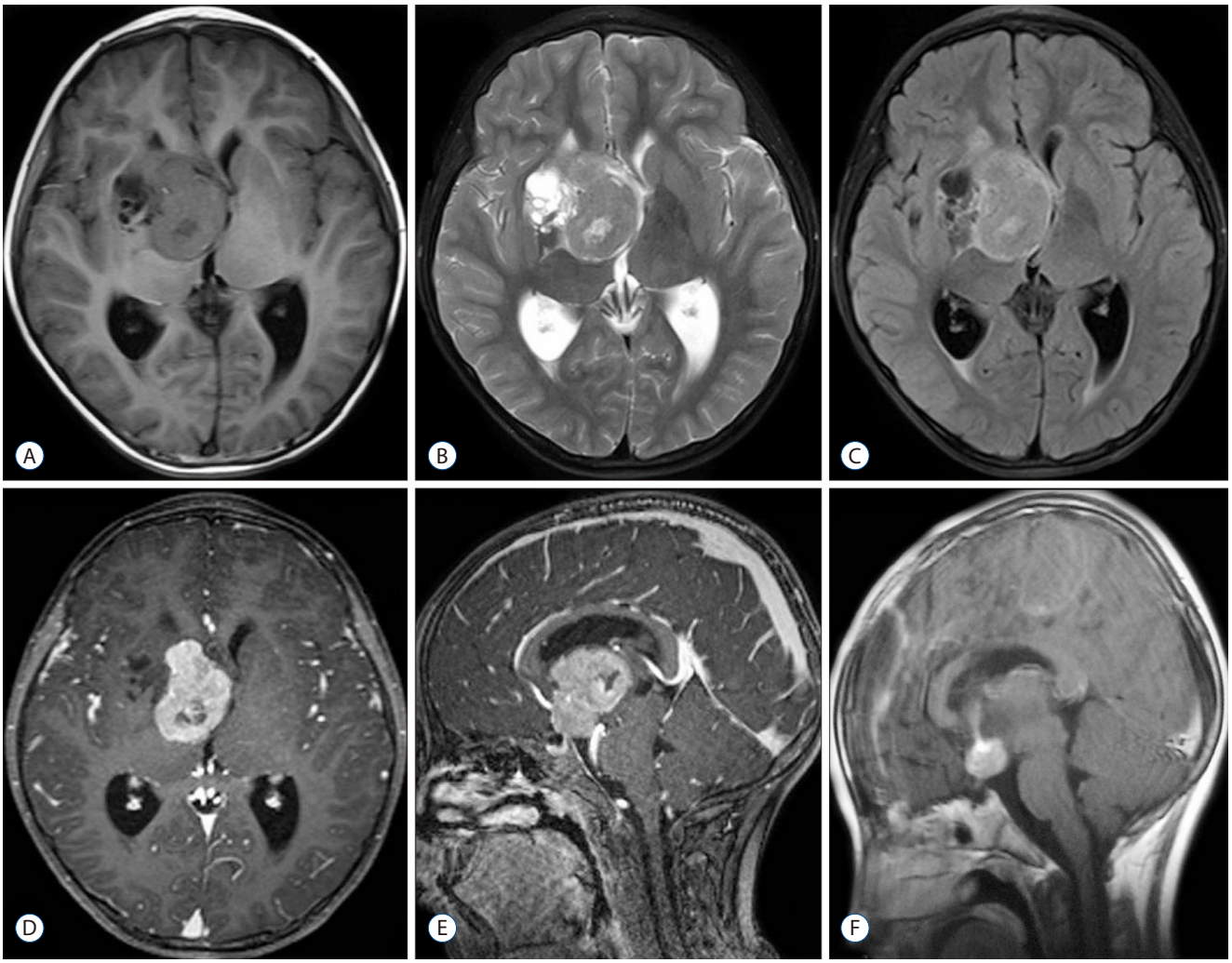

Fig. 2. MRI showing a mass lesion in the right basal ganglia consisted of solid and cystic parts. The solid part manifesting isointense signal on T1-weighted (A), T2-weighted (B), and FLAIR (C) with heterogeneous enhancement (D and E); the cystic part showing hypointense signal on both T1-weighted (A) and FLAIR (C) and hyperintensity on T2-weighted images (B) without enhancement of the wall (D and E). Although the lower margin of the tumor touched the dorsum sellae dura, 'dura tail' sign was not noted (E). Postoperative MRI revealing that most of the tumor has been resected and the residual part detached from the dorsum sellae (F). Subdural fluid accumulation in the surgical region was also detected (F). FLAIR : fluid attenuation inversion recovery, MRI : magetic resonance imaging.

al.[14,16] showed that extent of initial surgical resection was the strongest independent prognostic factor for pediatric meningiomas and upfront radiotherapy achieved no benefit. Hence, GTR was thetreatment of choice. In our case, STR was performed because of the rich blood supply, tight adhesion with ACA and lower tolerance of blood loss in children.

Zhang et al.[34] and Starshak[30] treated patients with malignant meningiomas with postoperative radiotherapy. However, postoperative radiotherapy was controversial. Some people thought meningioma could be induced by radiation[23]. Others suggested that adjuvant radiotherapy might delay recurrence of malignant meningiomas or progression of residual meningiomas[24]. Our case received postoperative radiotherapy because of the atypical meningioma and residual tumor growing.

Intraparenchymalmeningiomas were generally benignin children (n=11, 64.7%). Intraparenchymal atypical meningioma was first showed in zhang's study[2]. In our patient, the lesion also proved to be atypical meningioma (World Health Organization-II).

The prognosis is worse in pediatric than in adult population, depending on the degree of excision, pathologic grade,

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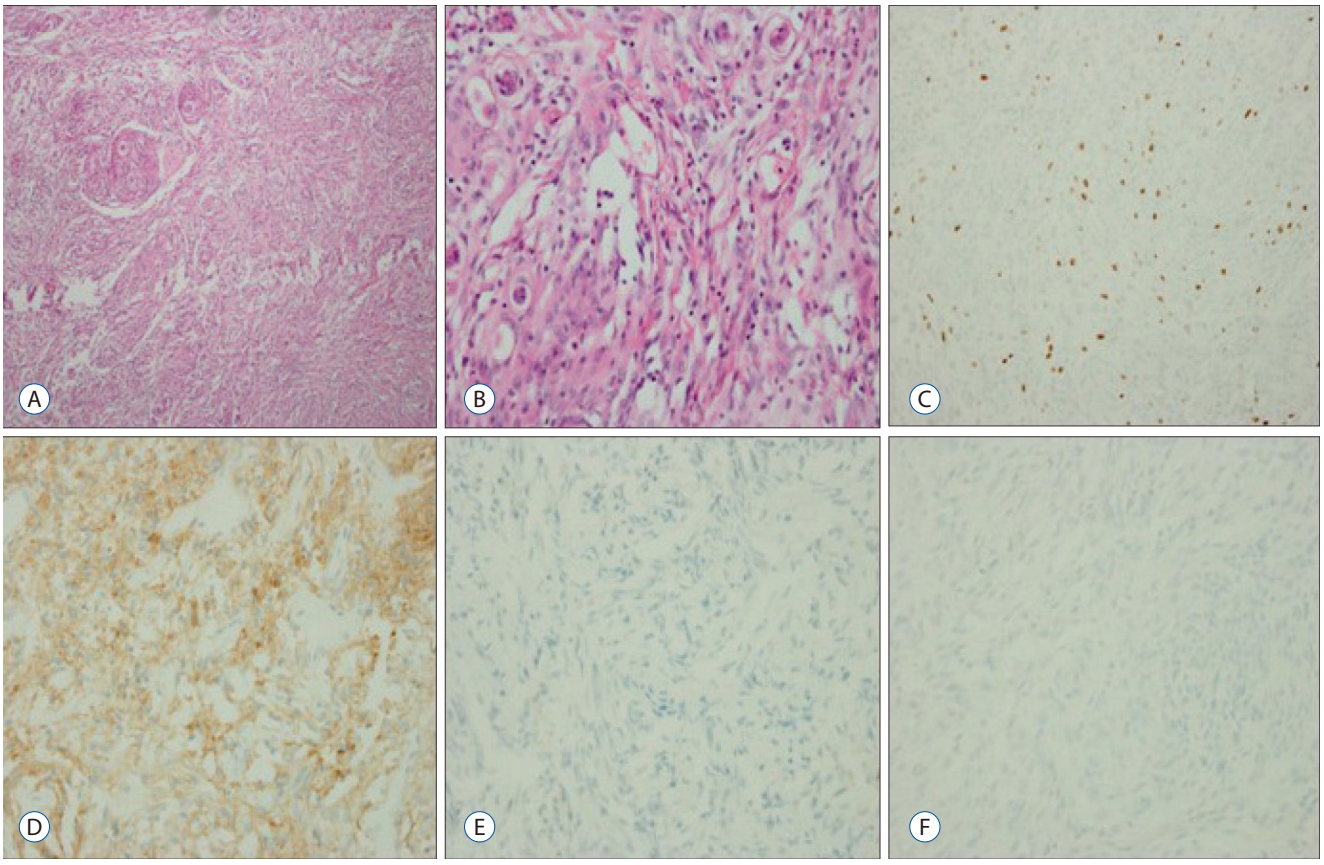

Fig. 3. Hematoxylin-eosin staining exhibiting sheetlike growth, foci of spontaneous, increased cellularity, small cells with a high nucleus-to-cytoplasm ratio and prominent nucleoli (A : original magnification, ×100; B : original magnification, ×400). Immunohistochemical staining showing positive for epitheliod membrane antigen (D : original magnification, ×400), but negative for glial fibrillary acidic protein (E : original magnification, ×400) and S-100 protein (F : original magnification, ×400). Ki-67 labeling index being approximately 10% (C : original magnification, ×400).

tumor location and association with neurofibromatosis[14,16,25]. The 10-year recurrence rate for GTR and STR is 33% and 82%, respectively[1]. Hence, Kotecha et al.[14,16] suggested the follow-up should be once every three months for at least 10 years and all life time for those with GTR or STR.

Features of radiology

The radiological features of pediatric primary intraparenchymal meningiomasinclude cystic component, calcification, large volumes and peritumoral edema. Compared with other meningiomas, the key difference of intraparenchymal meningiomas is the absence of dural attachment[10,27].

Differential diagnosis

The main radiological differential diagnosis for basal ganglia meningiomasincludes gliomas, lymphomas and germinomas, but sometimes it is quite difficult by imaging findings alone. Generally, low grade gliomas are usually iso- to hypodense on un-enhanced CT scans, whereas germinomas and lymphomas are of high density[5,12,21,24,28]. With progression and emergence of apredominantly solid component, it is more difficult to distinguish non-typical meningiomasfrom gliomas, germinomas or lymphomas, all of which are iso-dense to hyper-dense on un-enhanced CT scans and relatively isointense on all MR pulse sequences[5,12,21,24,28]. Cystic changes, intratumoral hemorrhage and heterogeneous enhancement are more frequently seen in germinomas and gliomas[24]. However, with the similar large size, peritumoral edema and mass effect are usually slighter in meningiomas than in gliomas. Hence, clinical manifestation and other supplementary examinations should be considered in such circumstances. For instance, it's helpful to identifygerminomas with tumor markers in serum and CSF or lymphoma with elevated lymphocytes percentage in peripheral blood and CSF[5,12,21,24,28]. Careful evaluation of

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Table 1. Summary of cases involving primary intraparenchymal meningiomas in the literature

Study	Age (years)/ Sex	Location	Clinical presenation	CT	MRI (solid part)			Surgery	Pathology	Postoperative treatment	Recurrence	Follow up
					T1	T2	CE					
Present case	8/M	Basal ganglia	Headach, vomiting, left hemiparesis	Iso/hyperdense, cyst, calcification	Isointense	Isointense	Heterogeneous	STR	Atypical	Rd	10% remnant	3 months
Nayil et al. (2015)[20]	3/M	Frontal	Headache, vomiting	NS	NS	NS	Heterogenous	GTR	Annplastic	No	No	NS
Werbrouck et al. (2014)[33]	13/M	Temporal	Seizure	Hyperdense, calcification	NS	Hypointense	Homogeneous	GTR	Fibrous	No	NS	NS
Jung and Song (2012)[8]	1.7/M	Frontoparietal	Seizure, hemiparesis	NS	Iso/hperintense	NS	Heterogeneous	GTR	Transitional	No	No	9 months
Pinto et al. (2012)[23]	17/F	Temporal	Seizure	NS	Hyperintense	Hypointense	Homogeneous	NS	NS	NS	NS	NS
Shimbo et al. (2011)[29]	10/M	Frontal	Seizure	Isodense	Iso/hypointense	Isointense	Homogeneous	GTR	Meningothelial	No	No	5 months
Zhang et al. (2007)[34]	16/M	Parietooccipital	Seizure	NS	Isointense	Hyperintense	Heterogeneous	GTR	Atypical	Rd	No	1.5 years
Karadereler et al. (2004)[10]	14/M	Temporal	Seizure, headache	NS	Hypointense	Hyperintense	Heterogeneous	GTR	Fibrous	No	No	3 years
Teo et al. (1998)[31]	1.8/F	brainstem	Hemiparesis	NS	NS	NS	NS	STR	Clear cell	Rd refused	70% remnant	NS
Starshak (1996)[30]	6.8/M	Frontal	Headache	NS	Heterogeneous	Heterogeneous	Heterogeneous	GTR	Sarcomatous	Rd, ch	No	5 years
Kohama et al. (1996)[13]	1.8/F	Frontal	Seizure	Hyperdense	Iso/hypointense	Hypointense	Homogeneous	GTR	Fibroblastic	No	No	2 years
Perilongo et al. (1992)[22]	2/M	Temporal	NS	NS	Isointense	NS	Homogeneous	GTR	NS	No	NS	NS
Mamourian et al. (1991)[18]	2/F	Frontal	Vomit, microcephaly	Heterogeneous, calcification	NS	NS	NS	GTR	Psammomatous	No	No	15 weeks
Schroeder et al. (1987)[27]	7/M	Frontal	Seizure	Hyperdense, calcification	Hypointense	Hypointense	NS	GTR	Fibroblastic	NS	NS	NS
Sakaki et al. (1987)[26]	0.9/M	Frontal	Seizure	NS	NS	NS	Homogeneous	GTR	Fibroblastic	No	No	5 years
Kimura et al. (1987)[11]	0.9/M	Frontal	Seizure	NS	NS	NS	NS	GTR	Fibrous	No	No	5 years
Drake et al. (1986)[2]	12/M	Temporal	Seizure	NS	NS	NS	NS	STR	Transitional	No	NS	3 years
Legius et al. (1985)[17]	1.2/M	Parietal	Seizure	Hyperdense	NS	NS	NS	GTR	Fibrous	No	NS	2.2 years
Morimoto et al. (1976)[19]	17/F	Parietal	Seizure	NS	NS	NS	NS	GTR	Anaplastic	NS	No	2.4 years

CE : contrast enhancement, M : male, STR : subtotal resection, NS : not stated, GTR : gross total resection, Rd : radiotherapy, F : female, ch : chemotherapy

radiologic features should be emphasized, which would assist in selection of the preferred treatment for patients.

Pathogenesis of primary intraparenchymal meningiomas

The pathogenesis of primary intraparenchymal meningiomas is unclear. Some theories are proposed to explain the possible mechanism : 1) intraparenchymal meningiomas arise from arachnoid cells of the piamater, which enter the brain along with perforating blood vessels[6,18,32,35]; 2) the meningiomas, which arise from the piamater of brain sulcus, adheres and compresses the brain parenchyma, and grows into the intraparenchymal lesion, so the mass is seen to be completely buried in the parenchyma[29]; 3) some authors presume that the arachnoid cells rest during the migration progress[6]; 4) intraparenchymal meningiomas are believed to arise from ectopic meningothelial cells within the stroma of the pia mater[6,18,32,35]; and 5) the occurrence may be due to cellular dedifferentiation within the cerebral parenchyma, or they may arise from the sheath cells of cranial nerves, which is proposed as the similar mechanism for the equally uncommon cutaneous meningioma and intradiploic meningioma[3]. The first two theories are the most probable mechanisms to explain the origin in our case, based on the close relationship with the perforating arteries of ACA and middle cerebral artery and recurrent artery of Heubner, the imaging features and intraoperative findings.

CONCLUSION

For lesions of basal ganglion extending to the superasellar region, lack of dural attachment is the key neuroimaging feature for differentiating intraparenchymal meningiomas from tuberculum sellae, clinoid, sphenoid wing and cavernous sinus meningiomas. Intraparenchymal meningiomas should be considered when gliomas, lymphomas, germinomas and other common lesions are excluded. The intraparenchymal atypical meningioma of basal ganglion in child is firstly reported, which should be emphasized in differential diagnosis to assist in selection of the preferred treatment for patient or avoiding a delay in management.

PATIENT CONSENT

The patient provided written informed consent for the publication and the use of their images.

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