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Case Report

Multiple myeloma presenting as dural plasmacytoma

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ABSTRACT

The finding of intracranial, extramedullary plasmacytoma is rare in multiple myeloma, especially with dural involvement. Meningioma remains the most common intracranial extra-axial mass. We report a case of a 39-year-old male who presented with intracranial, extra-axial mass found later to be dural plasmacytoma and additional multiple lesions on skeletal survey, leading to a diagnosis of multiple myeloma. The objective of this case is to increase awareness of the possibility of dural plasmacytoma as a differential diagnosis of meningioma and a harbinger of multiple myeloma. Magnetic resonance imaging plays a vital role in the detection of additional lesions in individuals while excluding multiple myeloma.

Introduction

Plasmacytoma is a plasma cell dyscrasia in which a plasma cell tumor grows in soft tissue or in the axial skeleton. There are 2 types of plasmacytomas: paraskeletal plasmacytomas consisting of soft-tissue masses arising from focal bone lesions and extramedullary plasmacytomas (EMPs) consisting of soft-tissue masses with no contact with bone [1]. Unlike the skeletal forms that frequently progress to multiple myeloma (MM) over the course of 2-4 years, the extramedullary forms rarely progress to MM [2]. The head and neck region are the most common location reported for extramedullary plasmacytoma [3]. The incidence of EMPs is 7% to 18% at MM

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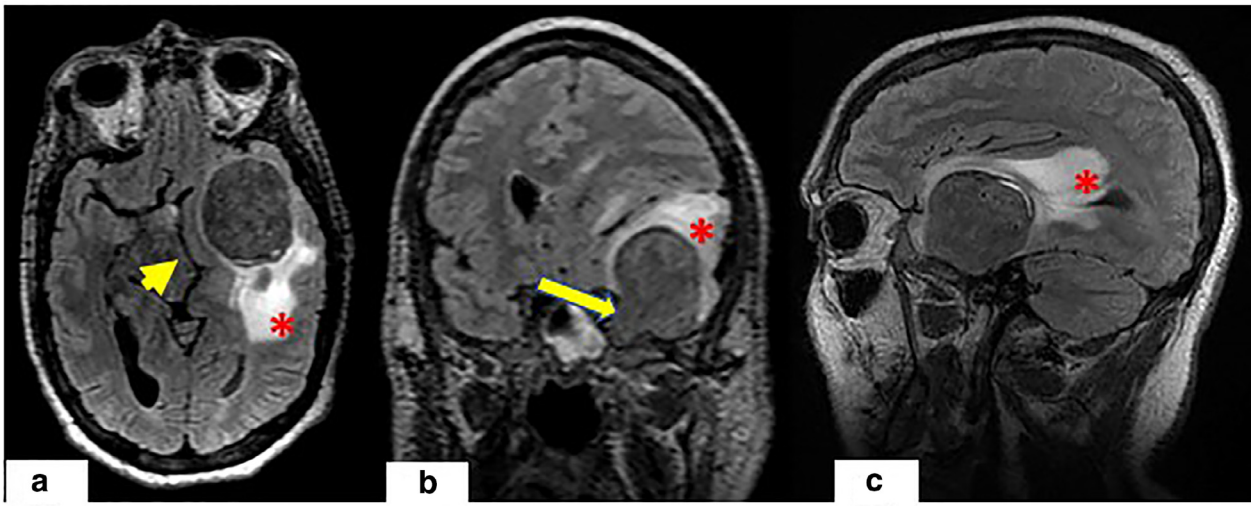

Fig. 1 – A 39-year-old male with dural plasmacytoma discovered to have multiple myeloma. Axial (1a), coronal (1b), and sagittal (1c) 3D fluid-attenuated inversion recovery (FLAIR) images showing a well-defined mass emerging from the left middle cranial fossa with surrounding vasogenic edema (asterisks) and mass effect. There is compression and displacement of the ipsilateral cerebral peduncle (short arrow), temporal lobe, and lateral ventricle with moderate midline shift and extension into the sphenoid sinus (long arrow).

diagnosis [4]. Etiology is unknown but might be related to differences in cellular adhesion molecules or chemokine receptor expression profiles of the malignant plasma cells [5]. EMPs account for approximately 3 percent of plasma cell malignancies. Approximately two-thirds of patients are male and the median age at diagnosis is 55 to 60 years [6,7]. The risk factor is unknown but the presence of plasmacytoma along with other factors such as male gender, age greater than 65 years, African American, and family history increases the risk for MM [5].

Case report

A 39-year-old man presented with 2 weeks history of worsening headache, gait abnormality, right facial weakness, and memory impairment. Notable medical history was testicular carcinoma 8 years prior, treated only by surgical resection. There was no history of urinary or fecal incontinence, limb weakness, or sensory deficits. A physical examination showed mild right facial weakness. Routine laboratory parameters were normal except for mild anemia (hemoglobin 11.2g/dL; ref 13.8-17.2 g/dL), hypocalcemia (serum calcium 8.3mg/dL; ref 8.5-10.5mg/dL), and elevated alkaline phosphatase (237 IU/L; ref 44-147 IU/L). Magnetic resonance of the brain without and with gadolinium showed a well-defined, partially lobulated, heterogeneously enhancing isointense left parasellar mass, measuring 5.0 × 4.4 cm, rising from the floor of the left middle cranial fossa (Figs. 1-3). This was favored to represent a meningioma.

Computed tomography (CT) angiography (Fig. 4) was obtained prior to stealth-guided pterional craniotomy and tumor resection. The mass was again noted as a well-circumscribed, homogenously enhancing hyperdense tumor (97 Hounsfield unit, HU) with displacement of the left posterior and middle cerebral arteries. Microscopic examination during tumor resection revealed a dural-based tumor with plasmacytoid features on histology with plasmacytoma and MM as the differential diagnosis (Fig. 5). Postoperative urine protein electrophoresis and immunotyping revealed albumin, alpha-1, alpha-2, beta, gamma components with minor restriction band in beta region and monoclonal kappa light chain. Serum protein electrophoresis and immunotyping showed elevated microglobulin levels including beta-2 (4.2 g/dL; ref 0.7-1.5 g/dL), alpha-2 (1.0 g/dL; ref 0.3-0.9 g/dL), and gamma globulin (1.7 g/dL; ref 0.5-1.4 g/dL), IgA (645 mg/dL; ref 80- 350 mg/dL) with a major restriction band in the gamma region.

MRI of the spine (Fig. 6) acquired during the same admission in a quest to rule out MM, revealed an enhancing epidural mass and multiple similar enhancing lesions scattered in the spinous processes, intervertebral foramen, L5 and sacral vertebrae, ribs, and extramedullary sites such as the posterior chest wall muscles. The patient underwent a T3 and T4 laminectomy and epidural mass resection. Microscopic examination was consistent again with a tumor of plasma cell origin and a diagnosis of MM. He was therefore referred to an oncology center for further treatment.

Discussion

Solitary dural plasmacytoma (SDP) is composed of plasma cells that are histologically and immunophenotypically identical to EMPs seen in MM; however, the treatment of these 2 entities differs significantly, necessitating a careful review of the diagnosis. A differentiation between these 2 entities is made based on the exclusion of additional lesions in patients with SDP. EMPs can be an early presentation of, or progress toward, MM. Intracranial locations of plasmacytoma vary from

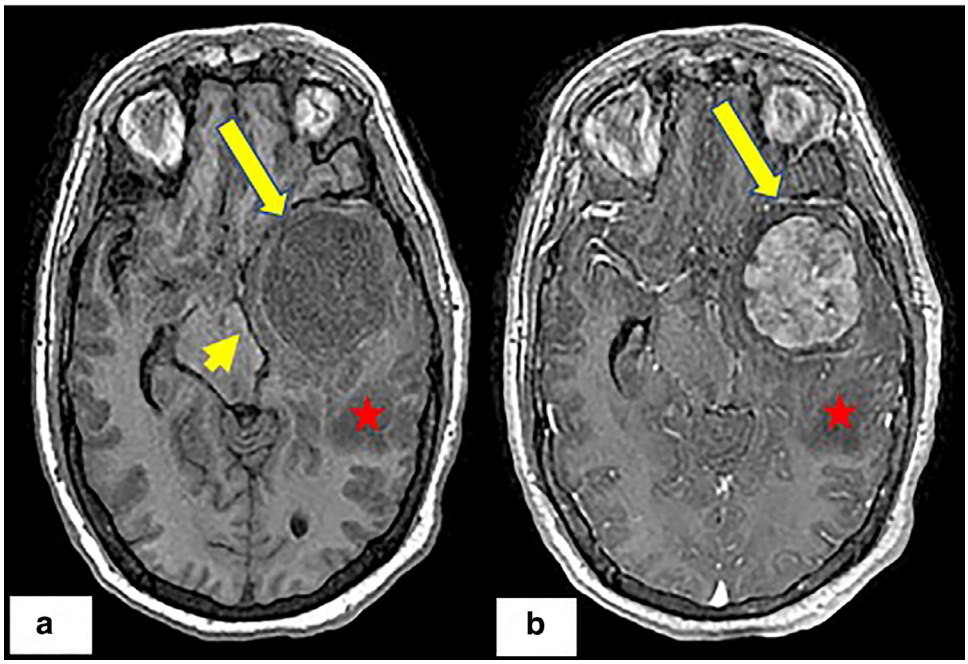

Fig. 2 – A 39-year-old male with dural plasmacytoma discovered to have multiple myeloma. T1-weighted (TW1) axial images without (2a) and with gadolinium (2b) show heterogeneous enhancement of the mass which was isointense to grey matter and perilesional edema (star). The left lateral sulcus, middle cerebral artery (long arrows), and cerebral peduncle (short arrow) are compressed and displaced.

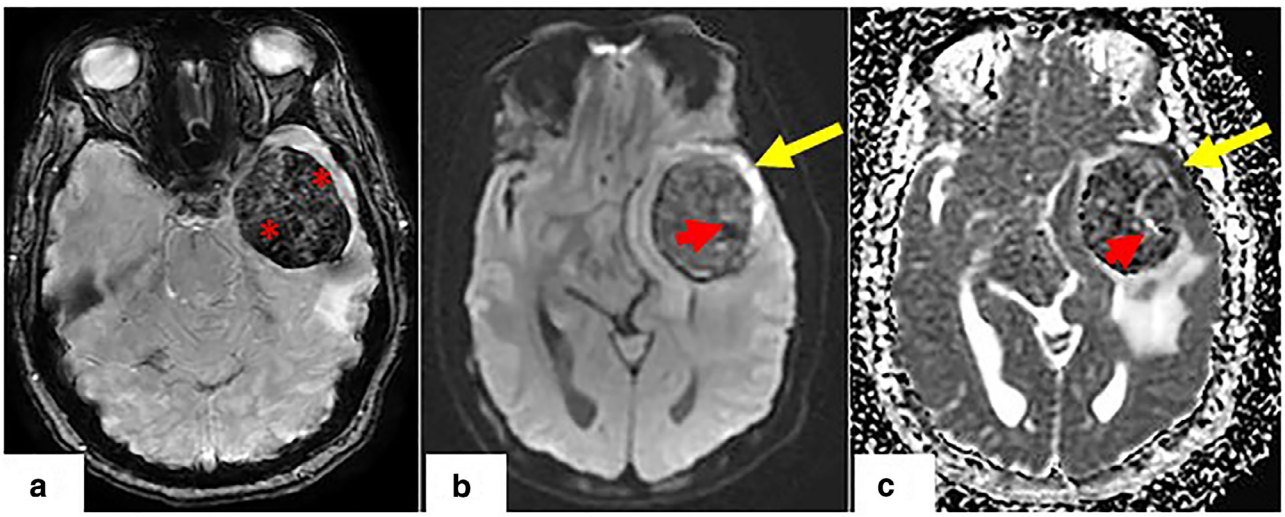

Fig. 3 – A 39-year-old male with dural plasmacytoma discovered to have multiple myeloma. (3a) Susceptibility-weighted axial image (SWI) shows dark signals (asterisks) suggestive of intratumoral hemorrhages. (3b) Diffusion weighted (DWI) and (3c) apparent diffusion coefficient (ADC) axial images show intratumoral hemorrhages (short arrows) as well as perilesional region of restricted diffusion (long arrows) to suggest ischemia.

cranial vault, skull base, sphenoclival, cavernous sinus, to diffuse or focal meningeal involvement [8–13]. Different locations of dural plasmacytoma such as parafalcine, frontotemporal, spinal, and cerebellopontine have been reported such that clinical presentation can mimic meningioma. Chronic headache, seizure, speech and gait disturbances, paresis, vertigo, hearing loss, dizziness mood changes, and other focal neurological symptoms have been reported [10,12–18].

The mass appears hyperdense on CT and enhances with contrast. Although variable appearances have been documented on MRI [19,20], the tumors are mostly high signal intensity on the T2W sequences but may be isointense,

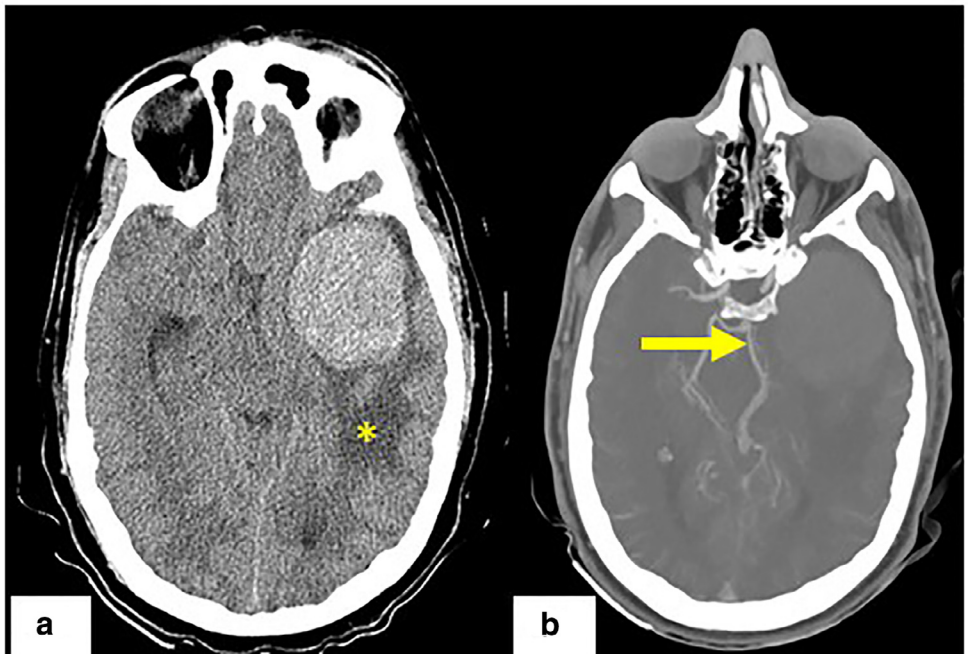

Fig. 4 – A 39-year-old male with dural plasmacytoma discovered to have multiple myeloma. Computed tomography noncontrast axial image (4a) show the mass to be hyperdense (97 HU suggestive of intratumoral bleed) with perilesional edema (asterisk). The angiographic axial image (4b) show displacement of the left posterior cerebral artery (arrow). The left middle cerebral artery is not seen in this plane.

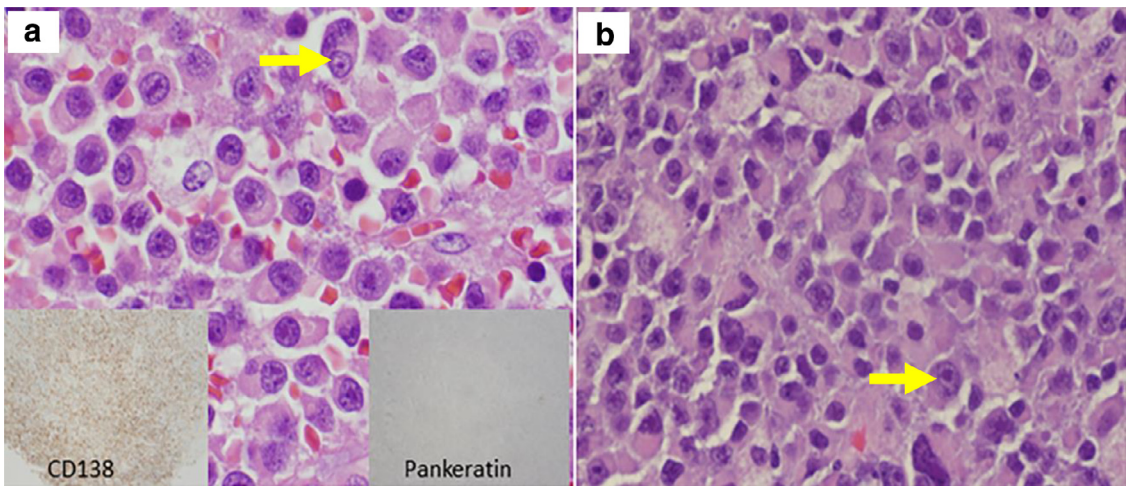

Fig. 5 – A 39-year-old male with dural plasmacytoma discovered to have multiple myeloma. Hematoxylin and Eosin staining x400 show neoplastic proliferation of differentiated plasma cells of the middle cranial fossa mass (5a) and epidural mass (5b). Note the "clock face" pattern of nuclear chromatin and perinuclear halo (arrows). Immunochemistry (inset) was positive for CD138, a plasma cell marker and negative for pankeratin (marker for other tumors like glial tumors, malignant meningioma, or metastasis).

isointense to slightly hypointense with respect to grey matter on T1-weighted sequences. Enhancement after administration of contrast can be homogenous or heterogenous therefore primary diagnosis based on imaging alone is usually not possible [19–21]. When dural plasmacytoma is pathologically found following surgical resection, postoperative systemic evaluation should include bone marrow examination, serum and urine protein electrophoresis, bone scan, and skeletal survey to exclude MM. An MRI of the spine has been reported to be a useful prognostic tool in asymptomatic MM revealing additional bone lesions in 50% with asymptomatic MM [22–24]. Whole-body MRI may be another technique to

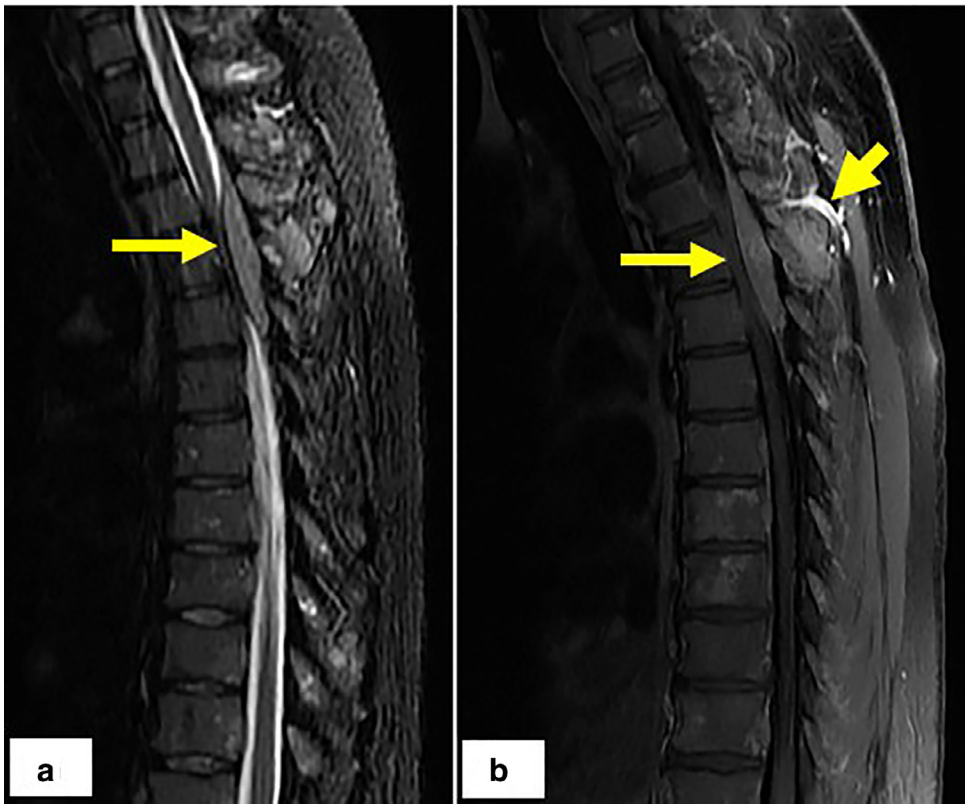

Fig. 6 – A 39-year-old male with dural plasmacytoma discovered to have multiple myeloma. MRI of the sagittal spine STIR (6a) and T1-weighted fat-saturated with gadolinium (6b) show an enhancing epidural mass, compressing the spinal cord (long arrows) at the T3 to T5 vertebral levels. Multiple similar enhancing lesions are demonstrated in few other vertebral bodies, spinous processes, and posterior wall muscles. Prominent vascularity is demonstrated in the posterior thoracic wall lesion (short arrow).

consider in staging an apparent solitary extramedullary plasmacytoma, because the detection of multiple, recurrent, or new lesions will influence the course of management.

Spinal cord compression from an extramedullary plasmacytoma should be suspected in patients with severe back pain, weakness, or paresthesia of the lower extremities, or bladder or bowel dysfunction or incontinence. MRI or CT myelography of the entire spine must be performed immediately if this complication is suspected. On MRI, extramedullary plasmacytoma is usually hypointense or isointense on T1-weighted images and isointense or hyperintense on T2-weighted images [19]. The absence of additional lesions or systemic evidence of MM will confine the diagnosis to a SDP.

There are multitudes of other dural-based masses. Due to the lack of specificity of the neuroradiological findings, we have listed a few such as meningioma, metastasis, and lymphoma that should be considered [13,25].

Meningiomas are the commonest extra-axial tumors. They are more common in women unlike the dural plasmacytoma. The atypical and malignant meningiomas are however slightly more common in males [26]. The small lesions can be incidental findings while larger lesions can present with headache, altered mental status, paresis, and focal neurological signs depending on the location and mass effect. They are highly vascularized tumors that avidly enhance postcontrast on imaging. A total of 70%-75% of meningiomas appear hyperdense on noncontrast CT while the rest are isodense. Calcifications are seen in about 20%. They are usually isointense to grey matter on T1W imaging, but some can be hypointense. They may appear iso- or hyperintense on T2 as well and show perilesional edema [26,27].

Extra-axial intracranial metastases to the skull, dura, leptomeninges may arise through several situations. Hematogenous spread to the meninges is the most frequent cause. Bone metastases extending intracranially and primary dural metastases show the characteristic biconvex shape, usually associated with brain displacement away from the inner table. They can be multiple or solitary and show variable appearance on imaging. They can be iso- or hypodense on CT with intense contrast enhancement. Like meningioma, metastases are usually iso- or hypointense on T1, hyperintense on T2, and may exhibit avid enhancement with variable amount of perilesional edema. Hemorrhage may alter the appearance [28]. Although this patient was previously treated for testicular cancer, metastasis to the brain is rare except if the primary was choriocarcinoma. The origin of the majority of dural metastatic tumors are the breast, lung, kidney, prostate, lymphoma, or a melanoma [25].

Dural lymphoma is a rare type of primary central nervous system lymphoma. They are usually the low-grade marginal

zone B-cell lymphoma and therefore have indolent courses with better prognosis. They occur more commonly in women compared to the parenchymal primary central nervous system lymphoma. Headache and seizure are also the most frequent symptoms and locations including the skull and spine have been reported. Due to high cellularity, these tumors are typically somewhat hyperdense on CT and enhance vividly. They are isodense to grey matter on T1W with vivid enhancement (usually homogeneous) and iso to hypodense on T2W. Perilesional edema is common in adjacent brain [29].

Although, MM with EMP and SDP have similar clinical/histopathological features, the treatment and prognosis differ. Treatment options for dural plasmacytoma are surgical resection of the plasmacytoma followed by adjuvant radiotherapy due to its high radio sensitivity. Surgical approach will depend on location and extent of the tumor. In addition to surgical resection, MM will require systemic treatment with chemotherapy. Plasmacytomas in patients with MM and have been associated with a poor prognosis while SDP is associated with long-term survival [12].

Teaching point

Dural plasmacytoma of SDP and from extramedullary manifestation of MM may present in a similar fashion clinically and radiologically. Histopathological diagnosis should prompt postoperative skeletal survey preferably with whole body MRI, bone marrow aspiration, and immunoelectrophoresis for exclusion of MM.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2019.05.026.

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