Extranodal Rosai-Dorfman Disease with Paranasal Sinuses and Intracranial Involvement: a case report

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Rosai-Dorfman disease (RDD) is a rare, benign pseudolymphatous condition, predominantly involving lymph nodes. Extranodal RDD presenting as metachronous nasal and intracranial masses has not previously been described. Herein, we reported such a case of RDD in a 49 year-old female who presented with nasal obstruction and subsequent headache. Pre-enhanced sinus CT showed diffuse polypoid masses in the nasal cavity without bony destruction. MRI demonstrated a strongly enhanced, infiltrative lesion filling the sinonasal cavity. The patient underwent surgical resection, and histopathology showed emperipolesis (lymphophagocytosis) and immuno-histochemistry showed S-100 positivity. The follow-up MRI one year later showed sinonasal tumor recurrence. In addition, it revealed a newly developed intracranial dura-based mass that exhibited hypointense foci on T2-weighted imaging. Recognition of these imaging findings is helpful in suggesting the diagnosis of RDD even in the absence of lymphadenopathy.

CASE REPORT

A 49-year-old woman is a HBV carrier and presented with nasal obstruction for two years. She received medical treatment as chronic sinusitis but in vain. Due to persistent nasal obstruction with purulent discharge, she was admitted to our hospital for further evaluation and treatment. Physical examination showed polypoid submucosal masses in the nasal cavity. There was no palpable lymph node or any cutaneous lesion. Sinus CT without contrast enhancement demonstrated diffuse polypoid masses filling the nasal cavity without bony destruction. MRI of the head and neck revealed infiltrative lesions filling the nasal cavity and extending to the bilateral osteomeatal complex with resultant obstructive sinusitis. The lesion showed intermediate signal
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Although intensity on T1-weighted images, hypointensity on T2-weighted images, and exhibited strong, homogeneous contrast enhancement after administration of contrast medium (Fig. 1a, b). Neither enlarged cervical lymph node nor intracranial space-taking lesion was seen. She underwent surgical resection of the nasal mass lesion, and histopathological examination revealed diffuse infiltration of pleomorphic inflammatory cells, including lymphocytes, plasma cells and histiocytes. Emperipolesis (lymphophagocytosis) was noted in some affected histiocytes which subsequently demonstrated positive S-100 protein on immuno-histochemical analysis (Fig. 2). These pathological features were characteristic of RDD.

One year after first admission, she complained of progressive headaches. The follow-up MRI showed an intracranial extra-axial dura-based mass in the right frontal region in addition to the residual infiltrative sino-nasal mass. The mass was well-defined and measured 1.7 cm in diameter (Fig. 3a, 3b). It was isointense on T1-weighted images and heterogeneous isointense to the white matter with tiny markedly hypointense foci on T2-weighted images. No obvious perifocal edema was evidenced. The mass exhibited strong and homogenous enhancement after administration of contrast medium. She underwent surgical resection of the right frontal intracranial mass. The histological pathology revealed diffuse infiltration of histiocytes with abundant cytoplasm and emperipolesis in the fibrous stroma. The histiocytes were positive for S-100 protein and negative for CD1a. Thus the right frontal lesion was confirmed as being intracranial RDD. For the treatment of recurrent sinonasal RDD, she received radiotherapy (4200 cGy/21 fractions). Chemotherapy or corticosteroid therapy was not instituted after considering the risk of exacerbation of chronic hepatitis B of the patient. The recurrent sinonasal RDD regressed completely after radiotherapy. Follow-up MRI showed no tumor recurrence in the following 2 years.

DISCUSSION

RDD was first described in 1969 as sinus histiocytosis with massive lymphadenopathy by Rosai and Dorfman [7]. The disease is rare, histiocytic

Figure 1. a, b. Axial T2-weighted MR images and post-contrast enhanced T1-weighted images shows an infiltrative hypointense mass lesion with solid contrast enhancement and associated obstructive sinusitis.
Extranodal Rosai-Dorfman disease has a distinctive histological appearance characterized by infiltration of lymphoplasmic cells and histiocytes with emperipolesis. The histiocytes in RDD are immunopositive for S-100 protein, CD68 and negative for CD1a [8]. The disease typically affects young adults with the mean age of 20.6 years. There is a slight male to female predominance (male: female = 1.4 : 1) [1]. RDD usually presents with massive, painless bilateral cervical lymphadenopathy and systemic symptoms of fever and body weight loss. Symptoms of extranodal RDD depend on the size and location of the lesion. Sinonasal RDD may present as nasal discharge or obstruction, epitaxis, pharyngitis and tonsillitis [9]. Regarding CNS RDD, the most frequent locations include the cerebral convexities, the parasagittal, suprasellar, cavernous sinuses, and the petroclival regions. Cephhalgia, seizure or cranial nerve deficit are the common symptoms of intracranial RDD [10]. In our patient with metachronous sinonasal and intracranial RDD, the clinical presentations were long-standing nasal obstruction, rhinorrhea and subsequent headache. To our knowledge, our case is the first report of metachronous sinonasal and intracranial RDD.

SINonasal RDD may present as an infiltrative submucosal lesion or a discrete pseudotumor without bony destruction [5]. As seen in our cases, the lesion typically appears as soft tissue masses filling the

**Figure 2.** Photomicrograph of a tissue sample showing infiltrations of plasma cells, lymphocytes and histiocytes. The histiocytes are large, pale and with emperipolesis (arrow).

**Figure 3.** Post contrast enhanced T1-weighted images (a) shows a well-circumscribed dura-based enhancing mass in the right frontal region. The residual infiltrative enhancing soft tissue in the nasal cavity was also noted. On T2-weighted images, (b) there is a markedly hypointense focus within the mass lesion.
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Sinonasal system with mild bony erosion of the contagious walls on CT. It is slightly hyperintense to the muscle on T1-weighted MR images, with intermediate signals on T2 weighted MR images and strong, homogeneous contrast enhancement on enhanced T1 weighted MR images [5, 9]. MRI is superior to CT in the delineation of the lesion extent and in the discrimination of tumor invasion from the obstructive sinus secretions. The differential diagnosis includes chronic inflammatory disease, lymphoma, squamous cell carcinoma, and sinusonal polyposis. Chronic inflammatory disease such as Wegener’s granulomatosis typically presented as noular sinonasal masses with low signal intensity on both T1 and T2 weighted MR images, and homogenous enhancement on enhanced T1 weighted MR images. Wegener’s granulomatosis commonly causes septal and non-septal bone destruction, and usually associated with orbital invasion. It also involves respiratory tract or kidneys. Absence of bony destruction mitigated against the diagnosis of Wegener’s granulomatosis and squamous cell carcinoma. Lack of tracheobronchial or renal lesion also makes the diagnosis of Wegener’s granuloma unlikely. The longstanding history of our patient lessened the possibility of lymphoma. The solid enhancement pattern of our case excluded sinonasal polyposis which typically showed mucosal enhancement only. On the other hand, typical intracranial RDD usually shows a well-circumscribed, dura-based mass lesion with strong contrast enhancement. Hypointense foci within the intracranial RDD on T2-weighted images have been observed in some reported cases [10-13]. All these findings were well demonstrated in the intracranial lesion of our case, suggestive of a metachronous intracranial RDD. Although meningioma, lymphoma, granulomatous disease (TB, sarcoidosis) and metastasis may produce the likewise appearances, it is more likely that the intracranial lesion was due to multiorgan involvement of the single disease entity rather than co- incidental occurrence of different disease entities. Indeed, involvement of both the sinonasal system and intracranial region in extranodal RDD has already been documented in a few cases [3-6]. Furthermore, presence of intralesional marked hypointense foci on T2-weighted image could add weight to the diagnosis of intracranial RDD. Nevertheless, pathology was still needed to confirm the diagnosis so that optimal treatment could be instituted.

The prognosis of RDD is variable. Most patients have an indolent course that is characterized by exacerbations and remissions. Half of the patients with systemic disease will experience spontaneously regression, while 17% of patients will have asymptomatic persistent adenopathy, and the other 17% will have residual symptoms for 5–10 years after onset [12]. Extra-nodal RDD tends to be chronic and relapsing [14], and two thirds of patients with sinonasal RDD have persistent or progressive disease [1]. Specific therapy to RDD is still inconclusive. Radiotherapy, chemotherapy and steroids used alone or in combination with variable responses have been reported [10, 14]. For intracranial RDD, surgical resection of the intracranial mass is most effective [10]. In our case, the sinonasal RDD exhibited local recurrence after surgery. Radiotherapy was then given to our patient and no more recurrent disease was noted then. On the other hand, our intracranial RDD could be totally eradicated with surgical resection alone, without any intracranial recurrence.

In summary, RDD is a rare idiopathic histiocytosis which typically involves lymph nodes. We report a case of extranodal RDD involving the sinonasal system and intracranial area that may pose a diagnostic challenge both for the clinicians and radiologists. Although definite diagnosis of RDD relies on the histopathologic examination, imaging findings including diffusely enhanced polypoid sinonasal masses without bony destruction as well as a strongly enhanced dura-based intracranial mass with hypointense foci may provide diagnostic clues preoperatively.

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REFERENCE

7. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy. A newly recognized benign clinicopathological entity. Archives of Pathology 1969; 87: 63-70


鼻竇腔及顱內之淋巴結外 Rosai-Dorfman disease：
病例報告

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Rosai-Dorfman disease（以下簡稱 RDD）是一種罕見的良性疾病，主要引起淋巴結腫
大，也可侵犯淋巴結以外的區域，淋巴結外的 RDD 以異時性的鼻腔和顱內腫塊為表現的病例
未曾被報導過。我們提出此一病例為 49 歲女性患有鼻塞而後產生頭痛，CT 顯示鼻腔內部腫塊
但未造成骨骼破壞，MR 影像則顯示鼻竇腔內腫塊有良好的顯影，病人在接受手術治療後病理
組織顯示有 emperipolesis（lymphophagocytosis），且免疫染色呈現 S-100 陽性反應，一年
後追蹤 MR 影像發現除了鼻腔之復發腫塊外，顱內亦出現貼著硬腦膜之腫塊，且該腫塊在 T2
加權影像下有低訊號的區域，即使病人沒有淋巴結腫大，辨識這些影像特徵將有助於該疾病的
診斷。