A new phenomenon of Rosai-Dorfman disease: half body stabbing pain accompanied by hypopharyngeal, laryngeal, and cervical lymph node lesions

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To the Editor: Rosai-Dorfman disease (RDD), also named sinus histiocytosis with massive lymphadenopathy (SHML), was first reported by Rosai and Dorfman in 1969.[1] Nodal RDD is usually characterized by massive lymphadenopathy; however, extranodal RDD is uncommon, its more severe fibrosis and fewer histiocytes in the lesion make it more difficult to diagnose than nodal RDD.[2] We herein describe a patient with both extranodal RDD in the hypopharynx, larynx, and cervical lymph nodes and a long history of half body stabbing pain that disappeared after surgical excision.

The 62-year-old female patient was referred to the Department of Otorhinolaryngology, Head & Neck Surgery, West China Hospital, Sichuan University with complaints of mild breathing difficulty, dysphagia to solid food, aspiration during drinking and hoarseness of voice for 3 months. Other symptoms included a stabbing pain in her left half body after she survived from a knife wound in a riot 8 years ago which could be relieved after everyday massage. Laryngscopy revealed a concealed left vocal cord, a narrowing of left piriform sinus, and a thickening of the left epiglottic fold with smooth mucosa covering. Enhanced computed tomography revealed that hypopharyngeal neoplasm had extended to the paraglottic space through the gap between the laryngeal plate and the arytenoid cartilage, the neoplasm had a clear margin in the larynx, and there was no obvious laryngeal cartilage destruction. A small lymph node between the submandibular gland and anterior border of the sternocleidomastoid muscle at the hyoid bone level was also observed [Figure 1A–1C]. Preoperative routine laboratory examination was normal.

The patient underwent surgical excision of the neoplasm under general anesthesia. To prevent postoperative aspiration, we adopted certain measures such as intended lateralization and mild rotation of the remaining laryngeal entrance. During the surgery, three lymph nodes were harvested. On the following day, the patient told her daughter the long-lasting stabbing pain at the left half body disappeared when the daughter intended to perform the routine massage. Histopathologic examination showed that one cervical lymph node out of the three presented hyperplasia of the sinuses with abundant histiocytic cells in the lumen, with pale nuclei, small nucleoli, and numerous intracytoplasmic-cell-predominant lymphocytes. Some plasma cells and neutrophils were also observed. However, in the other two lymph nodes, reactive hyperplasia was found. The neoplasm from the hypopharynx and larynx showed an irregular histiocytic and lymphoid cellular infiltration mixed with fibroblasts and xantomatous cells that were more frequently present between follicles than in the lymph node lesions. A panel of monoclonal antibodies was administered, including those against T cells (CD3, CD4, and CD8), B cells (CD20), histiocytes (CD68), and Langerhans cells (CD1a, S100). SHML cells were positive for CD68 and S-100 protein while CD1a was negative [Figure 1D–1H]. These pathologic results were consistent with RDD diagnosis. No medicine was prescribed for the patient after surgery and the 1-year close postoperative follow-up indicated that she was in good condition.

RDD is a rare proliferative histiocytic disorder of unknown etiology. Its possible association with immune disorders and virus infection has been reported. Extranodal RDD was very rare and no more than 30 cases of laryngeal RDD have ever been reported in literature. Hypopharyngeal RDD has not been documented in any medical literature before. As the hallmark of the disease, emperipolesis, the nondestructive phagocytosis of lymphocytes or erythrocytes, is normally required for diagnosis.

RDD is essentially benign but can be fatal when located in a special place such as airway. Nonsurgical treatment such

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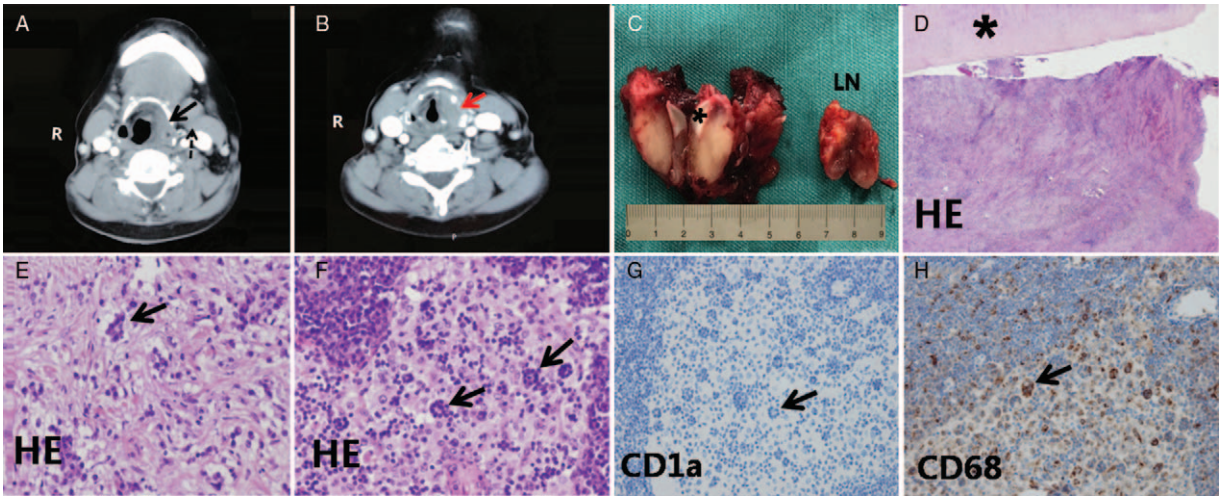

Figure 1: Computed tomography (A and B), operative specimen (C), and pathologic findings (D–H) of the patient. (A) Computed tomography demonstrated the circular thickening of the left hypopharyngeal mucosa (black arrow) and a lymph node (LN) enlargement (dashed black arrow). (B) The lesion in the hypopharynx extended to the paraglottic space (red arrow). (C) Operative specimen of the larynx and hypopharynx (left, *: cartilage) and LN. (D) Histologic examination revealed vaguely nodular polymorphous lymphohistiocytic infiltrate and no obvious destroy of the laryngeal cartilage (*) (hematoxylin-eosin staining, original magnification ×50). (E) In the neoplasm, histiocytes were focally clustered with numerous small mature lymphocytes and scattered plasma cells, emperipolesis was apparent (black arrow) (hematoxylin-eosin staining, original magnification ×400). (F) Emperipolesis was more apparent in the lymph node (black arrows) (hematoxylin-eosin staining, original magnification ×400). Sinus histiocytosis with massive lymphadenopathy cells were negative for CD1a (G, black arrow) and positive for CD68 (H, black arrow) (Immunochemical staining, original magnification ×400).

as steroids may be effective for the disease. For the lesions in the respiratory or digestive tract, surgery is the mainstay and sometimes may be urgently required.[3,4]

In brief, we report a nodal and an extremely rare case of extranodal hypopharyngeal and laryngeal RDD which may be probably relevant to trauma etiologically, and a new symptom – half body stabbing pain which has never been reported. In this case, it is difficult to explain the sudden disappearance of the pain and it remains a question whether it is relieved physiologically or psychologically.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the forms, the patient has given her consent for the use of patient's images and other clinical information in the journal. The patient understands that her names and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

Conflicts of interest

None.

References

1. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy: a newly recognized benign clinicopathologic entity. Arch Pathol 1969;87:63–70. doi:10.1016/j.pedex.2011.01.005.
2. Niu YY, Li YJ, Wang J, Jin XF, Yang DH, Huo H, *et al*. Laryngeal Rosai-Dorfman disease (sinus histiocytosis with massive lymphadenopathy): a retrospective study of 5 cases. Biomed Res Int 2017;2017:8521818. doi:10.1155/2017/8521818.
3. Aluffi P, Prestinari A, Ramponi A, Castri M, Pia F. Rosai-Dorfman disease of the larynx. J Laryngol Otol 2000;114:565–567. doi:10.1258/0022215001906192.
4. Bruce MW, Susan LA, Esther LC, Silloo BK, Dennis RH. Extranodal sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease) of the head and neck. Human Pathol 1993;24:483–492. doi:10.1016/0046-8177(93)90160-i.

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