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. Laryngoscopy 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 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 histiocyte 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 were 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. The neoplasm was positive for CD45 and negative for CD1a, CD31, CD21, and CD23. The neoplasm was also negative for CD1a and S-100 protein.[Figure 1A–IC]. 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 histiocyte 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.

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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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.

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