Original Article

Retrospective analysis of treatment strategies for nasal Rosai–Dorfman disease: Physiological perspective

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Abstract

Background and Aim: Rosai–Dorfman disease (RDD) is a rare form of lymphadenopathy with sinus histiocytosis, whose pathogenesis is undefined. Therefore, the purpose of this study was to explore the best treatment of RDD in the nasal cavity (NC), paranasal sinus (PS), and nasopharynx.

Methods: We conducted a review on the treatment stratigies published between 1969 and 2014 with duration of follow-up for nasal RDD along with 5 patients who received treatment in our hospital.

Results: The clinical data from literature review and combining with our 5 patients, we have 87 patients in total. There is a slight male to female predominance (male:Female = 1.81:1), average age was 37.36-year, the average duration of follow-up was 23.6-month. Lesions on 32 cases (36.8%) limited to the NC or PSs and 37 cases (42.5%) with involvement of other extranodal sites and 34 cases (40.2%) with lymphadenopathy (one unknown). 62 patients received operations for their initial treatment, 30 (48.39%) patients with recurrence, 21 of the patients received enlarged resection, and 5 (23.81%) patients with recurrence; 41 patients received cytoreductive surgery, and 25 (60.98%) patients recurrence; 20 cases received simple medical treatment, 16 (80.00%) patients with no treatment effect and 1 patients died. 5 patients recieved no therapy, survived with the disease with no progress.

Conclusion: Therapeutic effect of medical treatment was was not reported adequately in literature. Enlarged resection was the best way to treat nasal RDD. Enlarged resection combining with medical treatment could reduce the rate of recurrence, but this theory is just from few case reports and literature review. Finally, we found there was no relation between recurrence rate with age, sex, lymphadenopathy and other extranodal sites.

Key words: Medical treatment, nasal, recurrence, Rosai–Dorfman disease, surgery

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INTRODUCTION

Rosai–Dorfman disease (RDD) is also known as sinus histiocytosis with massive lymphadenopathy. The disease was first described by Destombes in 1965,[1] and later, described by Rosai and Dorfman in 1969.[2] RDD involves the cervical region in about 90%[3,4] of patients and every organ system can be affected and in 43%[5] of the cases the patients have at least one site of extranodal involvement, and 75% of them occurred within the head and neck region.[5] Within the head and neck, nasal cavity (NC) (50%) and paranasal sinuses (PSs) (18.7%) are the most affected sites followed by the salivary gland, oral cavity, pharynx and tonsils.[6] On the other hand, the most common symptoms in patients with nasal RDD (NC, PS, nasopharynx) are nasal obstruction, rhinitis, epistaxis, anosphrasia, tinnitus, ear stuff even hearing loss and physical findings are tissues fix to the NC, PS, nasopharynx. Computed tomography (CT) and magnetic resonance imaging most often reveal significant NC, nasopharynx, and PSs and often have polypoid masses, mucosal thickening, or soft tissue opacification. The diagnosis of RDD is according to the pathology. On immunostaining RDD show positivity for S–100 protein, CD68, CD14, CD33, and CD11c antigens and negative for CD1a.[7]

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Rosai–Dorfman disease is rare, and its pathogenesis is undefined. Though many etiologies of RDD have been

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postulated, such as infectious causes, immunodeficiency, autoimmune disease, and a neoplastic process, gene mutation,[8] and rhinoscleroma,[9] none has been substantiated. Hence, there was no definitive treatment for RDD. Nevertheless, multiple strategies of therapy have been used, including surgery, radiotherapy, chemotherapy, steroids, and so on. Currently, most reports of RDD addressed a single case, and larger case studies were rare and there were no clear records about treatment outcome especially for nasal lesions.

In this report, we retrospectively analyzed the treatment outcomes of 83 patients about nasal RDD from literature and also combining with 5 patients from our hospital who were diagnosed with RDD involving nasal region.

MATERIALS AND METHODS

From our database, we retrospectively studied the treatment outcome of 5 patients with RDD, who received enlarged resection by nasal endoscope in the Department of Otorhinolaryngology, Head and Neck Surgery of West China Hospital of Sichuan University and 83 patients diagnosed with nasal RDD in 29 English articles (keywords: RDD, NC, nasopharynx, PS) and 14 Chinese lectures from 1969 to 2014 involving nasal, NC, PS, all the 43 lectures contained the information of patients such as age, sex, treatment, prognosis, follow-up, lymphadenopathy, nasal diseased region, and other extranodal sites. Then we analyzed the above information with SPSS (Statistic Package for Social Science) 17.0 statistical software (SPSS Software Inc., Chicago, IL, USA) and also, calculated the recurrence rate with Kaplan–Meier ($P = 0.05$).

Case 1

A 45-year-old woman presented with a 7-year history of gradual onset of left nasal obstruction, left eyeball proptosis with limited outward and ingression, vision loss. Physical examination showed left middle nasal meatus filled with pink, soft neoplasm. No enlarged lymph node was found. Other physical examination, chest radiography, electrocardiogram, blood counts, erythrocyte sedimentation rate (ESR), coagulation, liver and renal function tests showed no abnormalities. The CT of nose and neck showed that the tumor mass occupied left nasal septum, left NC and maxillary sinus. Then, endoscopic sinus surgery of the mass was performed under general anesthesia. The operation revealed that left nasal septum and middle nasal meatus is obstructed by soft, fleshy, pink-white mass, bleeding not easily with manipulation. The masses and 3–5 mm safety margins of mass were completely removed by endoscopic surgery and the nasal biopsies were found to be diagnostic of RDD showing positivity for S–100 protein, CD68 antigen and negative for CD1a. Five-year of follow-up, showed no recurrence, and we did not find any lymphadenopathy.

Case 2

A 61-year-old woman presented with a 4-month history of snots flowed into mouth from postnaris with some blood steak, right nasal obstruction, and ear stuffy. Physical examination with the nasal speculum showed the NC, nasal septum, and inferior nasal concha was normal. No enlarged lymph node was found. Nasal endoscopic examination revealed right nasopharynx swelled and pressed homolateral pharyngeal orifice of eustachian tube. Nasal CT revealed a mass filled the right nasopharynx, and soft tissue in homolateral maxillary sinus, and some changes observed in the right mastoid portion. Then, complete resection of the neoplasm with nasal endoscopic surgery was performed under general anesthesia. The operation revealed tough, fleshy, smooth, white-yellow mass fixed to the right nasopharynx and filled maxillary sinus. The neoplasms were completely removed and the nasal biopsies were found to be diagnostic of RDD showing positivity for S–100 protein, CD68 antigen and negative for CD1a. Four-year of follow-up revealed no recurrence.

Case 3

A 47-year-old man presented with a Three-month history of gradual nasal obstruction, tinnitus and hearing loss. Endoscopic examination showed the right nasopharynx swelled with smooth, pink swelling without bleeding. There were no enlarged neck lymph nodes, cranial nerve deficits, or other anomalies in the head and neck region. Laboratory tests were normal. By enhanced magnetic resonance imaging, neoplasm was found in right parapharyngeal space, that extended to skull the base, oppressed nearby tissues, but the boundary was not clear. Complete resection of the neoplasm with nasal endoscopic surgery was performed under general anesthesia. The operation revealed tough, fleshy, smooth, white-yellow mass fixed to the right nasopharynx and filled maxillary sinus. The neoplasms were completely removed and the nasal biopsies were found to be diagnostic of RDD showing positivity for S–100 protein, CD68 antigen and negative for CD1a. 3 years of follow-up and there was no recurrence and enlarged lymph node.

Case 4

A 35-year-old man presented with a gradually developed left nasal obstruction, rhinorrhea, dizzy, headache, hyposmia since 1987, which were diagnosed as chronic sinusitis and left nasal polyposis. In 1997, he received a nasal endoscopic surgery for nasal polyp, and 3 years later, the symptoms recurred again. He received the second nasal endoscopic surgery. Postoperative, clinicopathologic diagnosis was rhinoscleroma. 1 year

later, he again felt the gradual nasal obstruction and intermittent nose bleed. In 2011, the man complained nasal obstruction, rhinorrhea, dizzy, headache and hyposmia. Physical examination with rhinoscope showed the left NC filled with pink-white, tough tissue, with slight bleeding, without peripheral lymphadenopathy or organomegaly. Laboratory tests showed that evaluated ESR 60 mm/h. Nose CT detected maxillary sinus, ethmoidal cellules, sphenoid sinus filled with soft tissues. There was also erosion of the medial walls of the maxillary sinuses, ethmoidal cellules, sphenoid sinus the septum, and parts of the ethmoid septae. Then complete resection of the neoplasm was done by nasal endoscopic surgery performed under general anesthesia. The operation revealed tough, fleshy, white-yellow mass fixed to the maxillary sinuses, ethmoidal cellules, sphenoid sinus the septum, and parts of the ethmoid septae. The neoplasms were removed and the nasal biopsies were found to be diagnostic of RDD showing positivity for S-100 (+), CD68 (+), KP1 (+), PGM1 (+), CD1a (−), Acid Fast Bacilli (−), W.S(−). In 3 years of follow-up, there was recurrence presenting intermittent nasal bleeding, and the endoscopy showed bilateral NC covered with a mass of clots. After removing the clots, we could see the neoplasm with bleeding filling the cavity.

Case 5

A 49-year-old woman presented with a 4-month history of gradual onset of bilateral nasal obstruction, hyposmia, pus and bloodstains in snots, ear stuffy and hearing loss, low-grade fever and weight loss. Physical examination showed bilateral NC blocked by neoplasm, and the structures in the cavity could not be investigated. Physical examination revealed a smooth surfaced, painless, nontender, nonmatted 1 cm × 3 cm, semimobile subskin mass at the right submandibular and a 3 cm × 2 cm at the left jugulodigastric region. CT scan of nose and PSs revealed soft tissue opacification of the right cavity and maxillary sinus, and multiple enlarged lymphs were found in superior mediastinum, bilateral submaxillary space, side of carotid sheath and bilateral roots of neck. Histopathology from submandibular lymph nodes showed RDD. Blood tests revealed a normal complete blood count. The histiocytes showed an expression of CD68 and S100 protein but did not express CD1a, epithelial membrane antigen nor parvovirus B19 antigens. Four weeks after surgery, the patient received radiotherapy.

RESULTS

The clinical data about lecture review are summarized in Table 1 along with our 5 patients. We analyzed the age, sex, treatment, duration of follow-up, lymphadenopathy, and other extranodal sites. There is a slight male to female predominance (male: Female = 1.81:1), average age was 37.36-year, the average duration of follow-up was 23.6-month. Lesions on 32 cases (36.8%) limited to the NC or PSs and 37 cases (42.5%) with involvement of other extranodal sites and 34 cases (40.2%) with lymphadenopathy (one unknown). Sixty two patients received operations for their initial treatment, 30 (48.39%) patients with recurrence, 21 patients received enlarged resection, and 5 (23.81%) patients had recurrence; 41 patients received cytoreductive surgery (CS), and 25 (60.98%) patients faced recurrence. Twenty cases received medical treatment (containing hormonotherapy, radiotherapy, chemotherapy, comprehensive treatment without surgery), 16 patients (80.00%) with no treatment or outcome (recurrence, no-change, progress),1 patients died with the persistent RDD which eroded the kidney and lead to hemolytic uremic syndrome. Five patients received no therapy, but all of them survived with disease and with no progress. We found the recurrence rate was related only to treatment ($P < 0.05$), but not with age, sex, lymphadenopathy, nasal diseased region, and other extranodal sites ($P > 0.05$).

DISCUSSION

Results of the present study suggest that the origin of RDD is unclear and there are many postulated causes such as infectious causes, immunodeficiency, autoimmune disease, neoplastic process, gene mutation,[8] and rhinoscleroma.[9] However, none of the causes has been substaintiated till date. Hence, the optimum treatment has not been instituted, and multiple strategies of therapy have been used. Even some people thought RDD was a self-limited disease, most patients will recover without any treatment or live with lesions for many years, even the whole life.[10] However for nasal lesion, it persists for a long time,[11] and the disease leads to life- or function-threatening obstruction like nasal obstruction, with repeated epistaxis. The mass could even spread from nasal cavities/PSs to intracranial sites.[12,13] Hence, treatment is necessary for nasal lesions to decompress the upper respiratory system, to open the PS, to stop epistaxis and to restrain spreading to intracranial sites or to relieve pressure in the orbits.

In addition to surgery, there were also good results after using radiotherapy, chemotherapy, steroids, thalidomide,[14] anti-CD20 monoclonal antibody rituximab,[15] platelet-derived growth factor-receptor b-inhibitors such as imatinib,[16] newer cytotoxic agents such as cladribine. (2-chlorodeoxyadenosine) and clofarabine.[17] But in our literature reviews, we found that 22 cases received medical treatment (hormonotherapy, radiotherapy, chemotherapy, comprehensive treatment without any surgery), 16 patients (72.7%) with no

Table 1: The summaries of RDD in nasal region

Sex/age	Treatment	Follow-up (month)	Outcome	Lymphadenopathy	Nasal region	Other extranodal region
Male/81	ES	24	No recurrence	No	NC	
Male/29	CS	48	Recurrence	No	NC	Subglottic, skin
Male/15	CS	7	Recurrence	Yes	NC	Subglottic
Male/18	CS	12	Recurrence	No	NC, PS	
Male/25	CS	6	No recurrence	Yes	NC	
Female/49	CS	24	Recurrence	Yes	NC, PS	Trachea
Male/42	HT	3	Progress	Yes	NP	
Male/20	ES	12	No recurrence	No	NC, PS	
Female/78	HT	6	No recurrence	No	NC	Subglottic
Male/64	NT	1	No change	Yes	NC	Subglottic
Female/41	NT	1.5	No change	No	NC	
Male/20	ES	6	No recurrence	No	NC	
Male/6	HT	3	No change	Yes	PS	
Male/27	CS	12	No recurrence	No	NC, PS	
Male/28	CS	16	No recurrence	No	NC, PS	
Male/53	CS	12	Recurrence	No	NC, PS	
Female/56	CS	6	No recurrence	No	NC, NP	Intracranial
Female/43	CS+HT	12	No recurrence	No	NC, PS	
Female/24	HT	4	No change	No	NC	Skin
Male/46	CS	48	Recurrence	No	NC, PS	
Male/38	HT	3	Recurrence	No	NC, PS	Knee
Male/54	ES	36	Recurrence	Yes	NC	
Male/18	CS	12	Recurrence	No	NC, PS	Knee, skin
Female/23	CS	204	No recurrence	No	NC	
Male/6	ES	2	Recurrence	No	NC, PS	
Female/42	ES+HT	24	No recurrence	Yes	NC, NP, PS	
Male/43	HT	3.5	No change	No	NC	Orbit
Male/36	ES	48	No recurrence	No	NC	Skin, anklew
Male/12	CS	18	No recurrence	Yes	NC, NP, PS	Eyelid
Male/20	CS	1	Recurrence	No	NC, PS	Eyelid, orbit
Male/29	ES+CT	24	No recurrence	No	NC, PS	Orbit
Male/10	HT	3	No change	Yes	NC, NB, PS	
Female/72	CS	3	Recurrence	Yes	NC	
Male/22	CS+HT	12	Recurrence	Yes	NC, PS	Skin
Male/22	CS	1	Recurrence	No	NC, PS	Eyelid
Male/12	CS	1	Recurrence	No	NC	Oribit
Female/48	CS	12	Recurrence	Yes	NC	Supraglottic, parotid gland
Female/64	ES	12	No recurrence	No	NC	
Female/16	CS+CT	12	Recurrence	No	PS	Oribit
Female/22	CS	6	Recurrence	Yes	NC, PS	Subglottic
Male/39	ES	12	No recurrence	Yes	NC	
Male/69	CS	36	Recurrence	No	NC	
Female/54	ES	12	No recurrence	No	NC, NP	
Male/57	ES	84	No recurrence	No	NC, PS	Kidney, scroful, spine
Male/11	HT	204	Died	Yes	NC	Kidney, pancreas, lungs
Female/19	HT+RT+CT	84	No change	Yes	NC	Eyelid
Male/53	CS	12	Recurrence	No	NC, PS	Sacral
Female/48	CS	6	No recurrence	Yes	NC, PS	
Male/69	HT	13	No recurrence	Yes	NC	Skin
Female/22	NT	9	No change	Yes	NC	Subglottic
Male/56	HT	36	Recurrence	No	NC, PS	
Male/42	CS	180	Recurrence	Yes	NC, NB, PS	Skin, eye
Male/16	HT	36	Recurrence	Yes	NC, NB	Skin, eyelid
Female/44	CS+HT	12	Recurrence	Yes	NC	Subglottic
Male/29	HT	12	Recurrence	No	NC, PS	Eye
Female/38	HT	6	Recurrence	Yes	NC	Skin
Male/72	CS	3	Recurrence	-	NC	
Female/43	CS	12	Recurrence	No	NC	
Female/48	CS	12	Recurrence	Yes	NC	Skin, kidney, etc
Male/51	HT	6	Progress	No	NC	
Female/62	HT+CT	3	No recurrence	No	NC	
Male/15	HT+CT+RT	1	No change	Yes	NC	

Contd...

Table 1: Condt...

Sex/age	Treatment	Follow-up (month)	Outcome	Lymphadenopathy	Nasal region	Other extranodal region
Female/45	EC	2	Recurrence	Yes	NC	
Female/12	NT	24	No change	Yes	PS	Orbit, lacrimal gland
Male/41	CS	9	Recurrence	Yes	PS	Skin
Male/31	CS+RT	24	Recurrence	No	NC, PS	Oribit
Male/28	CS+HT	12	No recurrence	No	NC, PS	
Male/42	ES	6	No recurrence	No	NC, PS	
Male/17	HT	6	No change	No	NP	
Male/20	CS	6	No recurrence	No	NC	Bone
Male/53	CS	12	Recurrence	No	NC	
Female/72	CS	20	No recurrence	No	NC	
Male/40	CS	62	No recurrence	No	NC	
Female/52	CS	52	No recurrence	No	NC, NB	
Male/22	CS+HT	4	No recurrence	Yes	NC	
Male/17	HT	6	No recurrence	Yes	NP	
Female/17	ES	3	No recurrence	No	NC, NP	
Male/31	ES	6	Recurrence	No	NC, PS	
Male/10	CS	12	No recurrence	Yes	NC, PS	
Female/34	NT	60	No change	Yes	NC	
Male/69	ES	12	No recurrence	No	NC, PS	
Male/29	HT	12	Recurrence	No	NC, PS	Eye
Female/45	ES	84	No recurrence	No	NC, PS	
Female/61	ES	48	No recurrence	No	NC, PS, NP	
Male/47	ES	36	No recurrence	No	NC, PS, NP	
Male/35	CS	36	Recurrence	No	NC, PS, NP	
Female/49	ES+RT	7	Recurrence	Yes	NC, PS	

RDD: Rosai-dorfman disease, RT: Radiotherapy, EC: Endometrial cancer, CT: Chemotherapy, ES: Enlarged surgery, CS: Cytoreductive surgery, HT: Hormonotherapy, NT: No treatment, NC: Nasal cavity, NP: Nasopharynx, PS: Paranasal sinus, NB: Nasal bone

treatment effect (recurrence, no-change, progress) and 1 patient died for lesion infiltrating the kidney. Zhu *et al*.[18] also suggested that radiotherapy and steroid treatment were not always the optimal treatment. From the Dr. Rosai's registry monitored, 21% were alive with no evidence of disease, 15% possibly had persistent disease, 54% had stable persistent disease, and 1% had progressive disease.[4] Analyzing from the Dr. Rosai's data, the patients who got good therapeutic response to medical treatment in some case reports belong to self-limited ones.

On the other hand, surgery was indicated as the best choice for dealing with the local disease, and it often resulted in long-term remission.[19] From the literature review, we observed two main surgical methods. The first one is just resection of the swelling in nasal region by cytoreduction surgery, and the second one is the whole tumor resection with removal of the surrounding critical structures, termed as enlarged surgery (ES). According to the literature review, we found that enlarged surgical resection would get a lower recurrence rate than cytoreduction surgery. Out of 41 patients received CS, 25 patients (60.98%) had recurrence, while for 21 patients received enlarged resection, just only 5 patients (23.81%) had recurrence. For our 5 patients, we completely removed the whole tumors and cleared the surrounding critical structures with 1–3 mm, and there were no recurrence with 1–5 years follow-up. But case 4 had operation 3 times, because the 1st time surgery was just to remove the swelling that lead to repeated recurrence. Apart for nasal lesion, other extranal lesions like abdominal lesions,[20] intracranial lesions,[21] cutaneous lesions, and even in lymph nodes lesions, enlarged operation was the best method of treatment.

For the nasal RDD, enlarged resection by endoscopic surgery was superior lateral rhinotomy. Nasal endoscopic surgery was a useful way and optimal treatment for nasal involvement. Easy for nursing, little trauma, good effect, shorter operative time, rapid postoperative recovery, leaving no scar on face and skull were the advantages of endoscopic sinus surgery. It further suggested that endoscopic surgery was the optimal treatment option for the nasal involvement by RDD as it can assure an improvement airway after the operation, minimal complications and relatively long period of remission. Ku *et al*.,[22] and El-Banhawy *et al*.,[23] reported two cases of RDD involving the NC were treated by endoscopic resection had no recurrence up to 1-year and 1.5-year after operation. Our all 5 patients received nasal endoscopic surgery, without recurrence. But if nasal endoscopic surgery could not remove all the lesions, open operation was needed and after all, total resection was the guarantee to no recurrence. Moreover, Belcadhi *et al.*, introduced CO_2 laser therapy with endoscopic surgery (ESS), that had good outcome with no recurrence.[24]

Because nasal region involvement by RDD was an uncommon entity, the diagnostic imaging findings mimic other common lesions such as nasal polyp, rhinoscleroma and inflammatory reactions. Thus, preoperative evaluation was a challenge and without biopsy the disease was commonly misdiagnosed. When without a definitive diagnosis, some surgeon just removed the tumor,[23,25] ignoring the lesions adhere to the surrounding, thus the recurrence was high. Hence, definitives diagnosis was the important guide for enlarged resection.

In our research, we found 4 (57.14%) patients had recurrence in 7 patients who received cytoreduction surgery combining with medical treatment and for them low dose steroids, radiotherapy and chemotherapy were considered.[10] There was no statistical significance with sample CS (61.76%) ($P > 0.05$). Especially, there was one recurrence (33.33%) in the 3 patients received ES combined with medical treatment. The recurrence one was treated with radiotherapy, no recurrence in subjects with steroids and chemotherapy. Our research was different from many other reports.[18,26-28] We have studied not only in nasal region,[18] but also from other places. Surgery combining with medical treatment has good result with 1–7 years follow-up, as there was no recurrence.[27-29] However, from the above data, we found that only ES could reduce the recurrence rate. For recurrence, no relationship was established between involvement of extranodal sites and lymph nodes.

Limitations of the study

Though this study is a retrospective analysis of data, it is a randomized controlled trial (RCT). The cases we analyzed in this article have a higher positive treatment results. In the future, we will do the RCT to assess the best treatment reqiured for nasal RDD.

CONCLUSION

According to the literature review, the therapeutic effect of medical treatment was not so good for RDD. Our results suggest that when anatomically feasible, enlarged excision by endoscopy could be the best treatment option for persistence or recurrence of exclusively RDD in the nasal region. Also, enlarged tumor resection combined with medical treatment could reduce the rate of recurrence, but this theory is just from some case reports and literature review, may be there is a need of RCT in larger group to confirm. Therefore, from the present study, we conclude there was no relation between recurrence and lymphadenopathy and other extranodal sites by systems analysis.

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