Rare disease

Tonsillar follicular lymphoma in a child

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Summary

Follicular lymphomas (FL) are among the most common non-Hodgkin's lymphoma (NHL) in adults. However, they are rare in children making up less than 3% of paediatric NHL cases. They occur most commonly in the head and neck region, lymph nodes or tonsils, with occasional extra-nodal occurrences. Distinction of FL from potentially clonal but, reactive follicular hyperplasia is important. We report a case of a 6-year-old male child presenting with night stridor since 6 months. Clinical examination revealed asymmetrical enlargement of the left tonsil. Routine left tonsillectomy was performed and the specimen was sent for histopathological examination. Diagnosis of follicular lymphoma was made on histopathological examination and further confirmed by immunohistochemistry.

BACKGROUND

Paediatric lymphomas are the third most common malignancy in children and account for 13% of all childhood cancers.^{1 2} Sixty per cent of these lymphomas are non-rapidly growing Hodgkin's lymphomas.² Non-Hodgkin's lymphoma (NHL) occurring in paediatric population commonly include lymphoblastic lymphoma, Burkitt lymphoma, anaplastic large cell lymphoma (ALCL) and diffuse large B-cell lymphoma (DLBCL). Follicular lymphomas (FL) is primarily a disease of adulthood with a median age of 59 years and is rare in paediatric patients.^{2 3} However, the most common sites in paediatric population are the tonsils and lymph nodes of the head and neck region.⁴

CASE PRESENTATION

A 6-year-old male child presented with night stridor of 6 months duration. Clinical examination revealed asymmetrical enlargement of the left tonsil. The rest of the pharynx, larynx and systemic examination revealed no abnormality. Routine tonsillectomy was performed and the specimen was sent for histopathological examination.

Microscopic examination revealed tonsillar tissue with intact squamous epithelial lining. The underlying lymphoid tissue showed complete effacement of the architecture by uniform sized follicles occupying almost the entire parenchyma (figure 1A,B). A well formed mantle zone was completely absent. The nodules were expansive,

Figure 1 (A) Microscopic view showing effacement of tonsillar architecture by a tumour composed of uniformly sized follicles with an ill defined mantle zone, bordered at places by retraction artefact (H&E, $\times 100$). (B) Another view of the tumour demonstrating the squamous epithelial lining of the tonsil (H and E, $\times 100$).

Figure 2 The neoplastic follicles are composed of a variable admixture of centroblasts and centrocytes (H&E, $\times 400$). *Inset* shows higher magnifications of the same (H&E, $\times 1000$).

irregular and coalescent in some areas, with very little uninvolved tissue left between them. These nodules comprised an admixture of centroblast with prominent nucleoli and variable component of cleaved centrocytes (figure 2). Abnormal mitotic figures and apoptotic cells were also seen. Furthermore, clinico-radiological and haematological investigations revealed no evidence of reticulo-endothelial disease elsewhere. Immunohistochemistry was consistent with the diagnosis of paediatric FL with the neoplastic cells being positive for CD20, CD10 and negative for CD3. CD10 positivity was also seen outside the neoplastic follicles. The Bcl-2 marker decorated only the mantle zones of reactive follicles

and was essentially negative within the germinal centres. Intense Bcl-6 reactivity was also seen both within and outside the follicles (figure 3A–D).

No postsurgical residual disease was identified on positron emission tomography (PET) scan and the child is presently kept under close observation and regular follow-up, given the highly curable nature of paediatric FL in the absence of residual disease.

OUTCOME AND FOLLOW-UP

The child is under close follow-up and so far there has been no recurrence or evidence of any residual disease.

DISCUSSION

Histopathological examination of routine tonsillectomy specimens to screen for occult malignancy is a common practice throughout the world. Disparity exists among hospitals with regard to processing adenotonsillectomy specimens.⁵ Many authors^{5–7} opine that histopathological evaluation of tonsils should be performed only on specimens with risk factors—commonest being tonsillar asymmetry and age. However, others⁸ hold a contrasting view and advocate histopathological assessment of all tonsillectomy specimens believing that though the incidence of cancer is small, it is significant. In our set-up, histopathological evaluation of all resected tonsils is performed on a routine basis so as not to skip any occult malignancy, as in the present case.

Follicular lymphoma is rare in the paediatric age group and constitutes about 1–2% of all paediatric lymphomas.² The most common subtype (32%) of NHL occurring in the paediatric age in India was found to be precursor

Figure 3 Immunohistochemical staining revealing positivity for Bcl-6 (A), CD10 (B) and CD19 (C). Consistent with most paediatric follicular lymphomas the Bcl-2 marker is negative (D).

T-cell lymphoblastic lymphoma (T-LL) in a workshop organised by the International Network of Cancer Treatment and Research.⁹ In this study, DLBCL, Burkitt and Burkitt-like lymphoma, ALCL and precursor B-cell lymphoblastic lymphoma constituted, 25%, 11%, 10% and 3%, respectively, of all the paediatric NHL.² FL is the most common type of NHL in adults in 20–30% of the cases and is extremely rare in the paediatric age group accounting for less than 6.5% of childhood lymphoma, with a median age range of 7.5–11.7 years.⁴

FL occurs most commonly in the tonsils or lymph nodes of the head and neck region, with occasional extra-nodal occurrence at a variety of sites, including the gastrointestinal tract, parotid gland, kidney, epididymis, skin and testis.¹⁰

Paediatric FL have been documented as single case reports with only a few collected series.⁴

Strict criteria must be used in the diagnosis of FLs in children, to differentiate them from the much more frequently occurring reactive follicular hyperplasia. Frizzera and Murphy¹¹ based their diagnosis of FL on the following architectural and cytological features: (1) Total or extensive replacement of the nodal structure; (2) even distribution of nodules throughout the node or lesion, as opposed to the predominantly cortical distribution of follicles in reactive hyperplasia; (3) crowding of nodules with little interposed lymphoid tissue; (4) overall uniformity in size and shape of nodules as compared with that seen in follicles of reactive hyperplasia; (5) paucity of reactive lymphoid cells (immunoblasts and plasma cells) in the interfollicular areas, strikingly demonstrable by immunoperoxidase staining and (6) the cytological composition of the nodules.

Winberg *et al*¹² found that the most helpful features were effacement of nodal architecture, the presence of cytologically atypical cells in the interfollicular areas identical to those in the neoplastic follicles and the monomorphous appearance of these cells in the follicles.

The number of histiocytes containing tingible bodies was found to be a reliable feature for distinguishing reactive from neoplastic follicular proliferations in both adults and children. An increase in the number of follicles per unit surface area may also serve as a helpful parameter for distinguishing reactive from malignant follicular proliferation.¹²

Paediatric FL have many features indistinguishable from those seen in adults but, demonstrate an increased proportion that are localised, lack Bcl-2 protein expression and translocations, and are grade 2–3 but still indolent and apparently curable.²

Learning points

- ▶ Follicular lymphomas (FLs) are rare lymphomas in childhood, with usually low grade and indolent course, usually at nodal or tonsillar sites in the head and neck region.
- ▶ Distinction from potentially clonal but reactive follicular hyperplasia is important.
- ▶ Understanding the full spectrum of B-cell neoplasia in children and its unique features is important for diagnostic, therapeutic and academic purposes.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Shad A**, Magrath I. Non-Hodgkin's lymphoma. *Paediatr Clin North Am* 1997;**44**:863–90.
2. **Agarwal S**, Das P, Karak AK, *et al*. Childhood follicular Non-Hodgkin's Lymphoma: a case report and review of literature. *Indian J Med Paediatr Oncol* 2008;**29**:36–9.
3. **Anon**. A clinical evaluation of the International Lymphoma Study Group Classification of non-Hodgkin's lymphoma. The non-Hodgkin lymphoma classification project. *Blood* 1997;**89**:3909–18.
4. **Agarwal R**, Wang J. Paediatric follicular lymphoma a rare clinicopathologic entity. *Arch Pathol Lab Med* 2009;**133**:142–6.
5. **Randall DA**, Martin PJ, Thompson LDR. Routine histologic examination is unnecessary for tonsillectomy or adenoidectomy. *Laryngoscope* 2007;**117**:1600–4.
6. **Beatty MM**, Funk GF, Karnell LH, *et al*. Risk factors for malignancy in adult tonsils. *Head Neck* 1998;**20**:399–403.
7. **Mohamad I**, Hassan S, Salim R. The routine histopathological examination of tonsillectomy specimens at Hospital Universiti Sains Malaysia—a retrospective study and its implications. *Malays J Med Sci* 2007;**14**:53–5.
8. **Oluswasanmi AF**, Wood SJ, Baldwin DL, *et al*. Malignancy in asymmetrical but otherwise normal palatine tonsils. *Ear Nose Throat J* 2006;**85**:661–3.
9. **Naresh KN**, Advani S, Adde M, *et al*. Report of an International Network of Cancer Treatment and Research workshop on non-Hodgkin's lymphoma in developing countries. *Blood Cells Mol Dis* 2004;**33**:330–7.
10. **Lorsbach RB**, Shay-Seymore D, Moore J, *et al*. Clinicopathologic analysis of follicular lymphoma occurring in children. *Blood* 2002;**99**:1959–64.
11. **Frizzera G**, Murphy SB. Follicular (nodular) lymphoma in childhood: a rare clinical-pathological entity: report of eight cases from four cancer centres. *Cancer* 1979;**44**:2218–35.
12. **Winberg CD**, Nathwani BN, Bearman RM, *et al*. Follicular (nodular) lymphoma during the first two decades of life: a clinicopathologic study of 12 patients. *Cancer* 1981;**48**:2223–35.

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