**CASE REPORT**

Interdigitating dendritic cell sarcoma of salivary gland associated lymphoid tissue not associated with HHV-8 or EBV infection

N Barwell, R Howatson, R Jackson, A Johnson, R F Jarrett, G Cook

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Interdigitating dendritic cell sarcoma (IDCS) is an extremely rare malignancy derived from professional antigen presenting cells. This report describes a case of IDCS arising in the salivary gland associated lymphoid tissue of the parotid gland of a 51 year woman, presenting with a painless neck swelling. Histologically, sheets of S100+/CD68+/CD45+/CD34+/CD1a+ spindle cells were surrounded by an inflammatory infiltrate with no evidence of B or T cell clonal proliferations. No evidence of either human herpesvirus 8 or Epstein-Barr virus could be detected by quantitative polymerase chain reaction in the tumour cells with serological evidence of previous Epstein-Barr virus infection. The patient remains well and disease free 24 months after presentation without specific treatment.

**PATHOLOGICAL FINDINGS**

The resected lymph node showed reactive changes only. The resected deep lobe of the parotid gland revealed a well circumscribed white lesion (25 × 1 × 12 mm) on sectioning. Histological examination demonstrated preserved lymphoid architecture, with reactive follicular and paracortical hyperplasia around an ill defined nodule, consisting of a proliferation of large spindle shaped cells with large oval/round nuclei, prominent nucleoli, and occasional mitotic figures, arranged in sheets with no specific architectural features (fig 2). There was a large reactive infiltrate consisting of macrophages, granulocytes, and occasional plasma cells. The spindle cells stained positively for S100, CD68, and CD45 (Dako, Ely, Cambridgeshire, UK) but did not stain with CD1a (Immunotech/Coulter, UK), CD34 (Serotec, Oxford, UK), CD3 (Dako), or CD20 (Dako). There was no demonstrable evidence of clonal B or T cell proliferation, as determined by polymerase chain reaction (PCR) assessment of immunoglobulin heavy (IgH) chain and T cell receptor (TCR) gene rearrangements.

Quantitative PCR using TaqMan® methodology was used to detect EBV and HHV-8 genomes, as described previously.

**DISCUSSION**

In our present case study we analysed a tumour arising in the salivary gland associated lymphoid tissue of the parotid gland that demonstrates no association with either EBV or HHV-8.

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**Abbreviations:** EBV, Epstein-Barr virus; FDC, follicular dendritic cell; HIV, human herpesvirus; IDC, interdigitating dendritic cell; IDCS, interdigitating dendritic cell sarcoma; IgH, immunoglobulin heavy chain; PCR, polymerase chain reaction; TCR, T cell receptor
median age, 51 years; range, 13–86), with a male pre-
dominance (male to female ratio 19:13). Most reported
cases of IDCS have presented in lymph nodes, although
extranodal primary sites include the small intestine, naso-
pharynx, testis, skin, tonsil, and spleen, with secondary
involvement of bone marrow, bone, liver, spleen, lung, ovary,
and skin being reported occasionally.26

Microscopically, the tumour cells are predominantly
pleomorphic in shape, although occasionally, spindle shaped
cells are seen. Immunohistochemical studies show that the
tumour cells are positive for S-100 protein, CD45, HLA-DR,
and CD68 but negative for B cell and T cell markers, CD30,
CD1a, and complement receptors.2 Ultrastructurally, the
tumour cells possess complex interdigitating cytoplasmic
dendritic processes, and Birbeck granules are absent. The
differential diagnosis includes undifferentiated metastatic
carcinoma, malignant and Langerhans’ cell histiocytosis,
anaplastic large cell lymphoma, primary and metastatic
sarcoma, and malignant melanoma. The diagnosis of any
one of these entities can rarely be reached from light
microscopy alone, and immunohistochemical and ultrastruc-
tural studies can contribute to discriminating between them.

Genetically, as illustrated in our case, there is no evidence of
clonal IgH or TCR gene rearrangements.

There was no evidence of the presence of human
herpesvirus 8 viral genomes

The aetiology of IDCS and the closely related FDC sarcoma
remains unclear and a viral pathogenesis has previously been
suggested.34 HHV-8 has been implicated in several malignant
conditions. HHV-8 DNA sequences have been demonstrated
in all the variants of multicentric Castleman’s disease and are

Figure 1  Extranodal interdigitating dendritic cell sarcoma. Sagital
computerised tomographic image at the level of the mandible
demonstrating a mass lesion in the inferior pole of the right parotid
gland (arrow).

Figure 2  (A, B) Histological examination of the resected inferior
pole of the right parotid gland. The central lymphoid area is replaced by a
proliferation of large spindle shaped
cells with large oval or round nuclei and
prominent nucleoli (haematoxylin and
eosin stain; original magnification:
(A), ×400; (B), ×1000). (C, D)
Immunohistochemical examination
revealed the spindle cells to be S100+/CD45+/CD16–/CD34+/CD3–/
CD20– (original magnification:
(C), ×400; (D), ×1000).

Take home messages

- We describe a rare case of interdigitating dendritic cell sarcoma arising in the salivary gland associated lymphoid tissue of the parotid gland of a 51 year woman
- There was no evidence of B or T cell clonal proliferation
- Human herpesvirus 8 and Epstein-Barr virus DNA could not be detected by quantitative polymerase chain reaction in the tumour cells with serological evidence of previous Epstein-Barr virus infection
- The patient remains well and disease free 24 months after presentation without specific treatment

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universally present in Kaposi's sarcoma.\textsuperscript{1,7,8} In multiple myeloma, HHV-8 was proposed to have a pathogenic role through infection of non-neoplastic bone marrow dendritic cells, although subsequent reports have not established a causal link between HHV-8 infection and plasma cell dyscrasias.\textsuperscript{9} We tested the hypothesis that HHV-8 may play a role in the pathogenesis of IDCS and, as demonstrated, there was no evidence of the presence of HHV-8 viral genomes.

These tumours may show a spectrum of biological behaviour, from low to high grade clinical courses with rapid progression to death in some patients. Follow up data, available for 26 patients in the literature, demonstrate a mean follow up time of 17.6 months: at six months, 10 patients had died from disease progression and two further patients had died of other causes. At the time of our case report, 14 patients were still alive, of whom eight were alive with disease and six were without disease. The median overall survival is 10 months. Response to treatment is generally poor and the most effective treatment and reliable prognostic factors remain unknown. The patient presented here had a more benign clinical course, with no evidence of disease recurrence at 24 months from presentation without specific treatment.

In conclusion, we present a case of IDCS arising in parotid gland associated lymphoid tissue characterised by the absence of clonal T and B cell proliferation that was not associated with HHV-8 infection.

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