



Langerhans cell sarcoma of the head and neck



James E.F. Howard^{a,*}, Liam Masterson^{a,b,1}, Raghav C. Dwivedi^{a,1}, Piyush Jani^a

^a Department of Otolaryngology, Head and Neck Surgery, Addenbrooke's Hospital, Cambridge CB2 0QQ, UK

^b Department of Pathology, Tennis Court Road, University of Cambridge, CB1 1QP, UK

Contents

1. Introduction	180
2. Methods	181
3. Results	181
3.1. Management of H&N cohort	182
3.2. Survival	184
4. Discussion	184
4.1. Risk factors and surface markers	184
4.2. Transdifferentiation	184
4.3. Outcomes	185
4.4. Survival	186
5. Conclusions	186
Conflict of interests	186
Funding	186
References	187
Biography	187

ARTICLE INFO

Article history:

Received 23 August 2015

Received in revised form

24 November 2015

Accepted 23 December 2015

Keywords:

Langerhans cell sarcoma

Head and neck

Prognosis

Management

ABSTRACT

Head and neck Langerhans cell sarcoma (HNLCS) is a rare malignant tumor carrying a poor prognosis. The aim of this work was to perform a systematic review of HNLCS cases, examine outcomes, and develop an evidence-based management algorithm. We performed a systematic literature search yielding 16 studies with 17 cases of HNLCS; 33 studies with 55 Non-HNLCS were used as a comparison.

Mean disease-specific survival was 20.5 months ($SE \pm 5.1$) for HNLCS versus 26.2 months ($SE \pm 4.2$) for non-HNLCS. There was no significant difference in disease-specific ($p = 0.768$) or disease-free survival ($p = 0.880$) between the two cohorts. There was a significant difference in both disease-specific ($p = 0.044$) and disease-free survival ($p = 0.001$) between local, locoregional and disseminated disease favoring more limited disease.

HNLCS appears to present later, with more disseminated disease. Surgery remains the mainstay of treatment of local disease, however clear margins do not guarantee clearance.

© 2015 Elsevier Ireland Ltd. All rights reserved.

1. Introduction

Head and neck (HN) sarcomas constitute a rare heterogeneous group of cancers that arise from the soft tissue or bony compo-

ment of this region. These tumors account for less than 1% of all head and neck malignancies and carry a relatively poor prognosis as compared to their counterparts at other sites. Rhabdomyosarcoma is the commonest head and neck soft tissue sarcoma in both the adult and paediatric population; the other common sarcomas being angiosarcoma, dermatofibrosarcoma, liposarcoma, ewings sarcoma and osteosarcoma (O'Sullivan and Gullane, 2009). Langerhans cell sarcoma (LCS) is a rare form of soft tissue sarcoma that similar to Langerhans cell histiocytosis, is currently believed to derive from myeloid precursors (Berres et al., 2014).

* Corresponding author at: Department of Otolaryngology, Head and Neck Surgery, Addenbrooke's Hospital, Cambridge University Teaching Hospitals NHS Foundation Trust, Cambridge CB2 0QQ, UK. Fax: +44 1223217559.

E-mail address: j.howard4@nhs.net (J.E.F. Howard).

¹ Equal contribution.

Langerhans cells are antigen-presenting cells found within the supra-basal region of mucous membranes, the dermis, lymph nodes and thymus gland and are members of the histiocyte system. This system comprises 2 cellular subsets: antigen-processing (phagocytic) cells and antigen-presenting (dendritic) cells. Dendritic cells consist of the follicular and inter-digitating dendritic cells found within lymph nodes, and Langerhans cells found in the epithelia.

Langerhans cells can be distinguished by their morphology (characteristic longitudinally grooved nuclei and with the presence of Birbeck granules) and immunohistochemical profile CD1a+ve, S100+ve, CD21–ve, CD35–ve and CD68–ve (Pileri et al., 2002).

Langerhans cell tumors are classified by the World Health Organisation (WHO) into Langerhans cell histiocytosis (LCH) and Langerhans cell sarcoma (Swerdlow et al., 2008). LCH is relatively benign in comparison to LCS, however rarely transformation to the latter has been reported (Lee et al., 2006). LCS displays features of malignancy including unregulated growth, local invasion and the ability to recur and metastasise. Cytologically LCS can be differentiated from LCH as it displays malignant features (e.g., cellular atypia and increased mitotic activity). These malignant features, in association with positive immunohistochemical staining (CD1a, CD207 [Langerin] and S-100 protein) are confirmatory of LCS (Swerdlow et al., 2008).

Demonstration of Birbeck granules on electron microscopy was formerly the gold standard for identifying the Langerhans cell type (Weiss et al., 2001), however it should be noted that this sign is not always present due to sample processing errors, artefact (Nakayama et al., 2010) or genetic mutation (Verdijk et al., 2005) and has been removed from the current criteria.

LCS involving the head and neck region (HNLCS) is a rare disease with only a handful of cases reported in the literature to date. As a result of this, and variations in nomenclature, there is a lack of evidence regarding its pathophysiology and the most appropriate management strategies (Howard et al., 2015). The aim of this work is to review all HNLCS cases published in the literature and to discern if there is any difference in survival when compared to the LCS of non-HN sites. An attempt has been made to develop an evidence based management algorithm based on our new proposed method of classifying LCS for better understanding and stratification of patients based on prognosis (Verdijk et al., 2005).

2. Methods

We carried out a systematic literature review on MEDLINE on 4th April 2015 with no date or language restrictions using the exploded Medical Subject Heading terms: (“langerhans cell sarcoma”[MeSH Terms] OR (“langerhans”[All Fields] AND “cell”[All Fields] AND “sarcoma”[All Fields]) OR “langerhans cell sarcoma”[All Fields]) AND (“Head Neck”[Journal] OR (“head”[All Fields] AND “neck”[All Fields]) OR “head neck”[All Fields]) OR (“mouth”[MeSH Terms] OR “mouth”[All Fields] OR (“oral”[All Fields] AND “cavity”[All Fields]) OR “oral cavity”[All Fields]) OR (“oropharynx”[MeSH Terms] OR “oropharynx”[All Fields]) OR (“tongue”[MeSH Terms] OR “tongue”[All Fields]) OR (“palatine tonsil”[MeSH Terms] OR (“palatine”[All Fields] AND “tonsil”[All Fields]) OR “palatine tonsil”[All Fields] OR “tonsil”[All Fields]) OR (“mouth mucosa”[MeSH Terms] OR (“mouth”[All Fields] AND “mucosa”[All Fields]) OR “mouth mucosa”[All Fields] OR (“buccal”[All Fields] AND “mucosa”[All Fields]) OR “buccal mucosa”[All Fields]) OR (“paranasal sinuses”[MeSH Terms] OR (“paranasal”[All Fields] AND “sinus”[All Fields]) OR “paranasal sinuses”[All Fields]) OR (“paranasal”[All Fields] AND “sinus”[All Fields]) OR “paranasal sinus”[All Fields]) OR (“nose”[MeSH Terms] OR “nose”[All Fields]) OR (“ear”[MeSH Terms] OR “ear”[All Fields]) OR (“skin”[MeSH Terms] OR “skin”[All Fields]) OR (“lymph nodes”[MeSH Terms] OR

(“lymph”[All Fields] AND “nodes”[All Fields]) OR “lymph nodes”[All Fields] OR (“lymph”[All Fields] AND “node”[All Fields]) OR “lymph node”[All Fields])) and variations there-of. The abstracts and titles were independently reviewed by three authors (JH, LM & RCD), and relevant full texts were retrieved. Article reference lists were examined for further cases to complete the literature search. Exclusion criteria included any articles without individual subject data, and non-English articles with insufficient detail in the abstract for data extraction.

Inclusion criteria included articles with individual subject data on outcome, follow-up period and treatment modalities. All available data were extracted along with any demographic, diagnostic and disease characteristic/case specific information. For the pooled analysis, cases with overlapping or insufficient data to perform survival analysis were excluded. Cases involving the head and neck at diagnosis were assigned to the HNLCS cohort. All remaining cases were used as the comparator (non-HNLCS). Disease extent was classified into local, loco-regional (single site with involvement of regional draining lymph nodes) and disseminated cohorts in order to aid better understanding of this rare tumor and stratify the management options which are not well defined (Howard et al., 2015). All statistical calculations were performed using SPSS Version 21 (Chicago, IL, USA). Rates of overall, disease-specific and disease free survival were estimated by means of the Kaplan–Meier method and were compared by the log-rank test. A univariable model was developed using Cox regression to investigate presence of local versus locoregional versus disseminated disease as prognostic indicators. Time dependent co-variants were investigated to identify concordance with the proportional hazards assumption.

3. Results

The abstracts for 292 articles were reviewed. Of these, 239 were excluded, including 2 whose full text was not in English with insufficient data in the abstract for inclusion. The references of the remaining 53 articles were reviewed yielding a further 8 papers. Sixty-one articles underwent full text review from which 12 were excluded (Fig. 1). Sixteen studies yielding 17 cases were finally included for the HNLCS cohort (Table 1) (Nakayama et al., 2010; Ben-Ezra et al., 1991; Bohn et al., 2007; Sumida et al., 2008; Yoshimi et al., 2008; Chang et al., 2013; Chung et al., 2013; Keklik et al., 2013; Luo et al., 2011; Muslimani et al., 2012; Park, 2012; SAGRANSKY et al., 2013; Valentin-Nogueras et al., 2013; Wang et al., 2013; Lee et al., 2014; Ma et al., 2014). Thirty-three studies reporting 55 cases of Non-HNLCS were used as a comparison cohort (Pileri et al., 2002; Lee et al., 2006; Ben-Ezra et al., 1991; Chung et al., 2013; SAGRANSKY et al., 2013; Wood et al., 1984; Bonetti et al., 1985; Delabie et al., 1991; Tani et al., 1992; Lauritzen et al., 1994; Itoh et al., 2001; Misery et al., 2003; Diaz-Sarrio et al., 2007; Ferringer et al., 2006; Julg et al., 2006; Kawase et al., 2005; Langfort et al., 2009; Lian et al., 2006; Ratei et al., 2010; Saribeyoglu, 2009; Uchida et al., 2008; Zhao et al., 2009; Yang et al., 2012; Au et al., 2013; Chen et al., 2013; FURMANCZYK et al., 2012; Kwong, 2014; Li et al., 2013; Shimizu et al., 2012; Wang et al., 2012; West et al., 2013; Xu et al., 2012; Zwerdling et al., 2014; Jimura et al., 2014; Wang et al., 2015).

Within the HN cohort, the male to female ratio was 1.125:1 and the median age at presentation was 56.5 years (inter-quartile range 45–69 years). At diagnosis 24% of cases had local disease, 35% loco-regional, and 41% disseminated disease. The most common site was nodal disease, present in 88% of cases, followed by the skin in 47% of cases. This was followed by sites within the upper aero-digestive tract in 35% of cases consisting of 3 cases with tonsillar disease, 2 with disease in the nasopharynx and 1 in the piriform sinus. The next most common sites were the lungs and spleen in 4 and 3 cases

Download English Version:

<https://daneshyari.com/en/article/6113557>

Download Persian Version:

<https://daneshyari.com/article/6113557>

[Daneshyari.com](https://daneshyari.com)