Kikuchi-Fujimoto disease with lateral neck localisation: a case report

Malattia di Kikuchi-Fujimoto: descrizione di un caso clinico a localizzazione latero-cervicale

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Introduction

Kikuchi-Fujimoto disease (KFD), a benign clinical form of necrotizing lymphadenitis of unknown aetiology, involves primarily females between 20-30 years of age, with a female:male ratio of 4:1.

It is somewhat difficult to estimate the frequency of the disease in Western Countries, since more than 50% of the cases described refer to patients in Asia and the first report in the literature, specialised in otorhinolaryngology, appeared in 1985.

As yet, KFD is not well known and not easy to evaluate as a potential cause of benign lymphadenopathy in the cervical area.

Although, it is worthwhile pointing out that the cervical lymph node system, even in a little known disease, is, without doubt, that more frequently involved; in fact, those cases characterised by generalised lymphadenopathy, hepatosplenomegaly, i.e., involving the extra-lymph node system, appear to be far less frequent.

Case report

A 22-year-old female patient came to our attention with a swelling localised in the right rear mandibular angle (level II) onset of which occurred 1 month prior to hospitalisation, followed 20 days later by the appearance of another swelling in the homolateral
supraclavicular area (level V). The rapid evolution of
the adenopathy, despite immediate treatment with an-
ti-inflammatory drugs (oral flurbiprofen), led the pri-
mary care physician to hospitalise the patient.
The physical examination revealed, in the absence of
other clinical signs of importance, a voluminous
swelling located in the right rear mandibular angle,
the edges of which were irregular. The mass was of
hard parenchymatous consistency, slightly painful
upon palpation, hypomobile in the deeper levels,
which had rapidly reached 6 cm in diameter, and a
lateral neck polyadenitis more pronounced on the
right, in correspondence to the supraclavicular area.
Blood tests, carried out, upon admission, revealed
only an increase in the Erythrocyte Sedimentation
Rate (ESR), whereas tests to detect antibodies
against Toxoplasma, Measles, Cytomegalovirus, Ep-
stein-Barr virus and HIV, as well as the tuberculin
skin test, gave negative results.
Contrast enhanced computed tomography (CT) of the
neck revealed the presence, in the bilateral lateral
neck area, of "numerous hyperdense lesions, some of
which colliquated, in part grouped together in a single
mass with polycyclic margins, characteristic of lym-
phadenopathies". The latter appeared to be 'more nu-
erous and voluminous in the right lateral neck site,
where they reached from the rear mandibular angle
region to the supraclavicular region’, thus confirming
the clinical finding. The radiologist referred to a diag-
nosis of 'suspected' lympho-proliferative disease'.
On the basis of clinical and instrumental findings, a
fine needle aspiration biopsy (FNAB) was per-
formed. Results failed to provide useful diagnostic
information and it was, therefore, necessary to per-
form a lymph node biopsy in the right lateral neck
area.
Histological examination of 4 adjacent lymph nodes
revealed a well-preserved general architecture, in

Fig. 1. Right lateral neck lymph node: areas of ischaemic
necrosis in paracortical site [●]. (Haematoxylin-eosin,
120 X).

Fig. 2. Higher magnification reveals presence of intra-
venous fibrin deposits in necrotic areas [●]. (Haema-
toxylin-eosin), 240 X).

Fig. 3. Necrotic area with abundant nuclear debris, indi-
cating karyorexis, and histiocytes [●]. No granulocytes.
Venule walls have thickened and intensely eosinophil ap-
pearance [●] (Haematoxylin-eosin, 480 X).

Fig. 4. Presence of cells with clear, nucleolated nucleus,
and abundant cytoplasm (plasmocytoid monocytes) with-
in an area of necrosis [●]. (Haematoxylin-eosin, 480 X).
which multifocal areas of necrosis with a central deposit of fibrin and abundant nuclear debris (Fig. 1) were visible in the paracortical area (Fig. 2). No granulocytes were present (Fig. 3). The necrotic areas were surrounded by marked proliferation of plasmacytoid monocytes (Fig. 4) which, at immunohistochemical examination, revealed weak positivity for CD43 and CD68; histiocytes positive to myeloperoxidase were also present.

The characteristic absence of polymorphonucleates, together with the presence of a proliferation of plasmacytoid monocytes mixed with the necrosis in the paracortical site, prompted the definition of the lesion as necrotising non-suppurative lymphadenitis due to Kikuchi-Fujimoto lymphadenitis. The patient, therefore, underwent anti-inflammatory (oral nimesulide) and antibiotic (piperacilline 2 g b.i.d. i.m.) treatment and was monitored until the symptoms disappeared, about 20 days later. It was possible, at 18 months’ follow-up, to exclude recurrence of the disease.

Discussion

KFD, a disease frequently found in Oriental countries, was first reported in the literature in 1972 in Japan. Tanaka et al. recently advanced the hypothesis that the higher incidence observed in Asians might be due to a genetic factor, corresponding to an allele of the histocompatibility HLA class II system detected with a statistically significant frequency in the DNA of Japanese patients presenting the disease. The aetiology still remains to be defined, however, the hypothesis advanced, so far, suggests that a viral agent may be involved. This would trigger a hyperimmune reaction resulting in polyclonal activation of T lymphocytes with a cytotoxic action. Albeit, recent studies have excluded the presence of a viral genome in the cells of lymph nodes involved in KFD. Thus, it has been suggested that the triggering factor could be a not well-defined super-antigen of a proteic nature which, binding to the T lymphocyte receptors, would determine activation.

The pathogenetic role of impaired function of the immune system is, moreover, supported by the possible association of KFD with systemic erythematous lupus or with other autoimmune diseases.

From a clinical point of view, the symptoms most frequently associated with lymphadenomegaly are asthenia, fever, sometimes a slight weight loss and, as far as concerns blood tests, neutropenia with lymphocytosis is often observed and, as in the case described here, an increase of ESR. These are, therefore, non specific, non pathognomonic symptoms which, together with the mode of onset which is common to various infectious and neoplastic diseases involving the lymph node system, account for the difficulties encountered in the diagnosis of KFD and stress the importance of differential diagnosis. In this respect, it is worthwhile stressing that adenopathy, in the cervical district, may be not only the site of metastases resulting from neoplastic lesions in the head and neck but also “early sentinels of neoplastic dissemination due to tumours situated in various, and sometimes distant, organs”.

Bearing in mind these considerations, it becomes clear that it is necessary, in order to proceed with an appropriate therapeutic approach, to rapidly reach a diagnosis and, thus, avoid underestimating the condition, since KFD may, even if only rarely, have fatal consequences.

Moreover, it has been reported that some patients presenting KFD have been submitted to chemotherapy following an erroneous diagnosis of lymphoma. From a diagnostic viewpoint, lymph node biopsy is mandatory, in our opinion, and thus from the historical findings, which may include immuno-histochemical investigation in cases that are difficult to interpret, whilst FNAB does not always provide reliable data.

Symptomatic treatment is carried out using corticosteroids or non-steroidal anti-inflammatory drugs. Prognosis, which is constantly favourable, is characterised by regression of lymphadenopathy within a few months, even if cases of recurrence have been reported after a considerable period of time.

References

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