

Unusual Bulky Presentation due to Missed Diagnosis of Hodgkin Lymphoma

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Rec date: March 06, 2015, Acc date: March 20, 2015, Pub date: April 03, 2015

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Case Report

A 44-year-old woman was admitted to the emergency room presenting a voluminous neck and axillary tumor associated with an upper left limb swelling and progressively severe dyspnea. At triage, the patient stated that the swelling had presented itself about three years earlier and that during this time she had never sought medical care, had self-prescribed steroid medication and had managed to camouflage the bulky mass to her relatives. Clinical examination showed a massive swelling with evidence of mediastinal syndrome (venous distension of the neck, upper limb oedema, compensatory collateral circles and shortness of breath) (Figure 1).



Figure 1: Venous distension of the neck, upper limb oedema, compensatory collateral circles and shortness of breath

Laboratory evaluation showed an increased neutrophil count ($18.5 \times 10^3/\mu\text{l}$) and lymphopenia ($0.45 \times 10^3/\mu\text{l}$), probably related to steroid abuse. Inflammatory markers were elevated, while LDH was only slightly increased (274 UI/l, UNL 225 UI/l). A total body, contrast-enhanced CT scan revealed bulky lymphadenopathy involving the left retro-mandibular, laterocervical and mediastinal districts (75×65 mm), as well as left axilla (80×80 and 90×120 mm) dislocating the local vascular structures. The mass also involved the left breast and resulted in severe compression of the left lung (Figure 2). Due to hemodynamic instability, only a core needle biopsy could be performed. Histopathological examination documented the presence

of Reed-Sternberg cells (CD30+, CD15-/+ , CD20+/-, PAX-5+, CD45-, CD3-, CD68-, ALK-), diagnostic of a classical sclero-nodular Hodgkin lymphoma (HL). After the first course of ABVD (doxorubicin, bleomycin, vinblastine, dacarbazine), the clinical conditions improved with reduced dyspnea and visible shrinking of the mass. Unfortunately, 20 days later, the patient developed left-arm cellulitis and pneumonia that rapidly precipitated into irreversible septic shock by multidrug-resistant *Acinetobacter baumannii*.

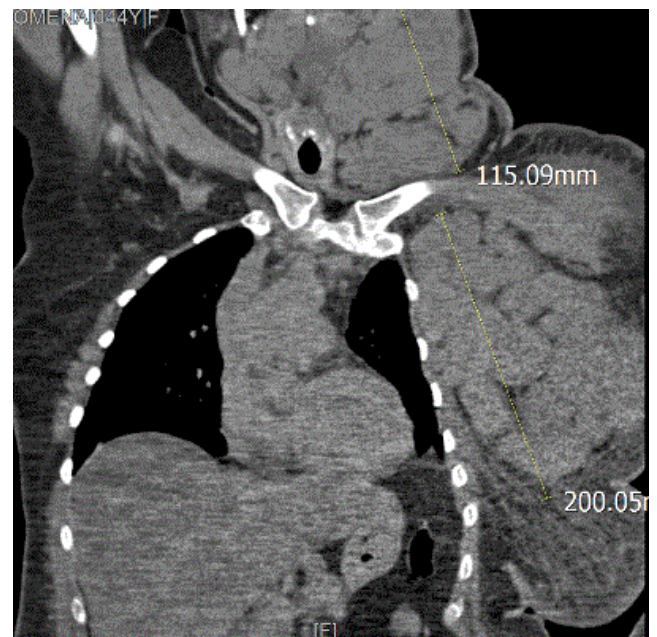


Figure 2: Mass also involved the left breast and resulted in severe compression of the left lung

HL accounts for approximately 0.6% of all new cancer cases in the USA. It is unique in that the neoplastic Hodgkin and Reed-Sternberg (HRS) cells of classical HL account for only 1% of the tumor tissue in most cases, with various inflammatory cells comprising the tumor microenvironment. Recent research has identified that HRS cells originate from germinal center B-cells [1].

In present days, characterized by early diagnosis and advanced medical techniques, the presentation of a 'visible' lymphoma with a 3-year delay appears rather remarkable, particularly in a country like Italy where the National Health System makes medical care available at no cost to all patients. The patient's habitus is reminiscent of the early days cases reported by Thomas Hodgkin in 1832, at a time when no effective treatment was available. At diagnosis, HL patients are

staged using the Ann Arbor System [2] and divided into 2 groups: those with limited-stage disease, typically including those with stage I or II disease (Involvement of 2 or more lymph node regions on the same side of the diaphragm); and those with advanced-stage (stage III or IV) disease.

Another interesting aspect of this case is that, despite the massive tumour mass and the years that elapsed from presentation to diagnosis, the lymphoma still had a limited stage II. To date, studies have established chemotherapy and involved-field radiotherapy (IFRT) as standard therapy in limited stages disease: 2 cycles of ABVD followed by 20 Gy for early favorable HL (according to the German Hodgkin Study Group criteria: No large mediastinal adenopathy, No Extra Nodal disease, 1-2 involved nodal regions, ESR \leq of 50 mm/hr) [3]; 4 cycles of ABVD plus 30 Gy for those with risk factors [4], like our patient. The outcome achieved by primary treatment with ABVD in major clinical trials and large single-institution series has improved over the years from the late 1980s through the early 2000s with a 5-year OS rate increased from just over 70% to approximately 90% [5].

In conclusion, it is likely that this woman lived with the stigma of having cancer, hiding her tumor and endangering her life, for a condition otherwise highly curable.

References

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