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Clinical case discussions

Extranodal lymphomas

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Extranodal or primary CNS DLBC lymphoma?

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A previously fit 58 year old male presented with a 6 month history of progressive sensory disturbance followed by motor weakness affecting both lower limbs such that he had lost the ability to walk. Upper limb and sphincter function was intact. There was no peripheral lymphadenopathy or hepatosplenomegaly. Neurological examination revealed mixed upper and lower motor neurone signs in keeping with a conus medullaris lesion. There was no apparent sensory level. Full blood count, biochemistry and LDH were normal. Imaging (CT and MRI) revealed a soft tissue mass filling the spinal canal from T11–L4, encasing the spinal cord, the conus and the cauda equine (image available). There was no evidence of disease elsewhere. A diagnosis of diffuse large B cell lymphoma was obtained following L3/4 laminectomy and biopsy. The tumour had a high proliferation index and immunohistochemistry revealed positivity for CD20, BCL6, BCL2 (weak), MUM1 and PAX5. The tumour was negative for CD138, CD5, CD10 and CD30. A lumbar puncture with CSF flow cytometry was not possible because of the location of the tumour mass. Bone marrow trephine showed no evidence of infiltration, in particular, no evidence of intravascular lymphoma. Our departmental clinical discussion focused around the optimal regimen to ensure adequate tumour penetration by chemotherapy. The consensus was to treat with R-CODOX-M/IVAC. Treatment is ongoing but the patient is showing prompt resolution of the neurological deficit and is now walking with assistance. Potential points for discussion: 1. Is this a primary CNS lymphoma? 2. What is the optimal management of a conus medullaris lymphoma? 3. Is there an association with intravascular B cell lymphoma? 4. Is there a role for consolidation radiotherapy?

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Intestinal T cell lymphoma

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A 59 year old female presented in June 2006 with sudden onset left iliac fossa pain on a background of chronic abdominal pain and weight loss. Clinical examination revealed features of an acute peritonitis. There was no palpable abdominal mass and no peripheral lymphadenopathy. She had a past medical history of coeliac disease. Full blood count and biochemistry including LDH were normal. CT scan showed free intraperitoneal gas, segmental small bowel thickening. She underwent laparotomy with jejunal resection and re-anastomosis. On histological examination there was extensive ulceration, necrosis and subtotal villous atrophy. The bowel wall was infiltrated with a dense, pleomorphic blast cell population which by immunohistochemistry was positive for CD2, CD3, CD7, CD30 and granzyme B. CD4, CD5 and CD8 were negative. Proliferation fraction was 60%. A diagnosis of enteropathy associated T cell lymphoma was made. CT post surgery was normal. Following 3 courses of CHOP, she had persistent abdominal pain and failed to gain weight and LDH was rising. CT scan did not show any small bowel abnormality. In view of the clinical suspicion regarding residual disease she proceeded to an MRI enteroclysis which showed diffuse thickening of the small bowel wall (image available). In view of the suboptimal response, treatment was changed to ESHAP, 3 courses, with stem cells harvested after course 2. Repeat MRI enteroclysis post ESHAP showed minimal residual nodular changes related to background coeliac disease but no residual mass (image available). She underwent a BEAM stem cell autograft in January 2007. She is currently well and in sustained remission three years post procedure. Potential points for discussion: 1. What is the optimal induction therapy for enteropathy associated T cell lymphoma? 2. Should all fit patients be considered for autologous transplant in first CR? 3. Should MRI be preferred to CT in assessing response to treatment?

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Primary hepatic low-grade B-cell lymphoma: A case report

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Introduction: Primary hepatic lymphoma is a rare disorder representing less than 1% of all extranodal lymphomas and there is no consensus on the best approach for management. Histological examination of a primary hepatic lymphoma usually reveals a diffuse large B-cell lymphoma; there have been few reports of primary hepatic mucosa-associated lymphoid tissue (MALT) lymphomas. We describe the case of low-grade B-cell lymphoma occurring in the liver.
Case report A: 74-year-old man with a history of a little pain on his right abdomen was presented in our hospital. He had no other complaints, no weight loss, fever or night sweats. Abdominal ultrasound showed a liver mass. A computed tomography (ct) scan of the abdomen was performed and it revealed a solitary mass (55×25×60 mm) in medial segment of the left lobe of the liver. Ultrasound-guided biopsy of the liver tumor suggested low-grade B-cell lymphoma Immunohistochemically, lymphoma cells were positive for CD20 and negative for CD5, CD10, CD3, CD43, Bcl-2. Staging procedures showed no lymphoma lesion other than the liver tumor, and all the biochemical blood tests were normal. Thus, the patient was diagnosed with low-grade hepatic marginal zone B-cell lymphoma. Considering that it was low-grade

lymphoma and the patient age he has been just followed up for the last 6 months. Blood cell count, serochemical findings including liver enzymes are normal and the liver mass is stable.

Discussion: Primary hepatic lymphoma is defined as lymphoma either confined to the liver or having major liver involvement. It represents less than 1% of all extranodal lymphomas. The exact cause of primary hepatic lymphoma is unknown, and there is no consensus on the best approach for management.

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Surgery and multiple successive chemotherapy helped to achieve a sustainable remission in patient with advanced MALT lymphoma

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Pt suffered from dramatic weight loss and acute pain, however, the CT did not revealed tumour and he was not referred to the oncologist. Only after suffering became unbearable he was admitted to the hospital with advanced lymphoma. Aggressive treatment improved conditions and remission was achieved. Pt E.A, 62 y.o. Diagnosis MALT lymphoma with the involvement of the small intestines and lymph nodes of mesentery with the secretion of the IgA lambda-type paraprotein, IVB stage Immunohistochemistry: proliferation of small lymphocytes with infiltration of intrafollicular structures. Phenotype: CD 20+, CD79+? CD5–, CD10–, CD23–, CD43+/–, Cd11+/–. In 1997 the pt noted the dramatic weight loss (14 kg), acute abdominal pain, vomiting. The level of the IgA lambda-type paraprotein was increased (15 g/l). CT did not detect tumour. In December 1999 due to intraperitoneal bleeding he underwent surgery with the resection of the small intestine, sanitisation and drainage of the abdominal cavity. Histological findings helped to set the diagnosis. February 2000 – January 2001 – 7 courses of chemotherapy (Leukeran, Vinblastin, Natulan, Prednisolonum). After that the pt was monitored. In December 2007 the increase of paraprotein (40 g/l) was observed. However the conditions remained stable, and interventions were not conducted. In February 2008 due to intoxication the chemotherapy (Vincristine, Carmustine, Alkeran, Cyclophosphan, Prednisolonum) was started. After 4 courses the pt had serious complications: granulocytopenia (4 grd) and the bilaterial mycotic pneumonia. New chemotherapy line (4 courses of Rituximab, Cyclophosphan, Fludara) started in December 2008 reduced intoxication symptoms. August – September 2009 – 2 administration of Rituximab to consolidate the response. From October 2009 – case monitoring. In December 2009 the paraprotein level was 0.9 g/l.

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A case report of myeloma in gastric MALT lymphoma with bone marrow(BM) involvement: Two distinct entities?

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Plasmocytic differentiation (PD) may been seen in a minority of B-cell non Hodgkin's lymphoma (NHL) but it's frequent in extranodal marginal zone B-cell lymphoma (MALT lymphoma). PD often manifests clinically with secretion on monoclonal immunoglobulin. In some cases, the distinction between a B-cell NHL with PD and a plasma cell neoplasm on the basis of immunohistochemical analysis on biopsy specimen may be challenging to diagnose. Recent data suggest that flow cytometric immunophenotyping (FCI) of plasma cell might help the differential diagnosis of these two entities. We report a case of MALT gastric lymphoma stage I that, after five years from diagnosis and treatment, showed BM involvement from nodal marginal zone B-cell NHL together gastric relapse, progressive increment of serum monoclonal component IgG-lambda and appearance of free light chain lambda on urine immunofixation. The patient received six cycles of R-CVP achieving a CR on gastric and BM biopsies specimens, but serum monoclonal component level was not affected and on the contrary it increased on. After 10 months BM biopsy detected an infiltration of 40% of small lymphoma cells stained with anti-CD20 monoclonal antibody associated with plasma cell differentiation. Rituximab $\times 4$ were performed, with further increase of serum monoclonal component IgG-lambda (4.9 gr/dl) and persistent BJ proteinuria. A new BM biopsy showed 90% of small lymphoid cell expressing CD20, CD79a, Bcl2 and lambda light chain. FCI of the BM was done and revealed an increased plasma cell population expressing CD38, CD138, CD20, CD56, CD117, but lacked CD 45 and cytoplasmatic light chain lambda. Further appropriate laboratory and radiographic studies defined the diagnosis of low-grade myeloma, small cell type, stage II (D–S) and stage III (ISS) and consistent treatment. FCI of plasma cell in correlation with the clinicopathologic data confirms its usefulness for the identification and characterization of myeloma cell.

Leukaemias

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Bone marrow necrosis: A challenging condition

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Bone marrow necrosis (BMN) is a rare entity, which is generally associated with malignancy. Hematologic malignancies are the underlying cause in almost 60% of the cases, with acute leukemia being the most frequent etiology accounting for 40%. Prognosis is generally poor but in some cases repopulation of the BM cavity with normal hematopoietic cells is seen. Treatment includes supportive measures and treatment of the underlying cause. A 54-year-old man with idiopathic myelofibrosis since 2004, treated with splenectomy and hidroxiurea, was asymptomatic until November/2009, when he presented with asthenia, anorexia and weight loss. Full blood count showed increasing basophiles but bone marrow aspiration didn't reveal leukemic transformation (LT). Two months later, he developed an intense and disabling pain on the lower back leading to hospitalization. Laboratory findings included Hg – 8.7 g/dl, white cell count – 8.07×10^9/L, platelets – 307×10^9/L, LDH – 1000 U/L (normal range: 67–190 U/L). During hospitalization, he developed pancytopenia and LDH raised up to 7354 U/L. Bone marrow (BM) aspiration and biopsy showed chronic myelofibrosis with extensive necrosis and no blasts. BM scanning revealed 99m Tc uptake only in proximal third of the left humerus, right humerus's head and both femur's heads. There was also an intense