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Myeloid sarcoma in non-leukemic and leukemic patients

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Μυελοειδές σάρκωμα σε μη λευχαιμικούς και λευχαιμικούς ασθενείς

Περίληψη στο τέλος του άρθρου

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Spinal myeloid sarcoma (SMS) is an uncommon entity associated with acute myeloid leukemia (AML) in up to 90% of the cases, or other myeloproliferative diseases; either preceding, coinciding, or following the diagnosis of these hematologic disturbances, and the confirmed diagnosis of people with isolated SMS has been very scarcely reported.[1–8] This tumor is equally cited as granulocytic sarcoma, myeloblastoma or chloroma, and affects the skin, lymph nodes, small intestine, and bones, but isolated spine involvement is rare.[1–8] The exceeding rare early onset of the tumor in infancy is related to genetic predisposition,[3] and the estimated incidence of isolated neoplasm in adults is 2 in 1 million population.[5] The consensual management schedule for patients with confirmed diagnosis of SMS has not been entirely established yet, and the current options include tumor surgical excision, chemotherapy, radiotherapy, besides a hematopoietic stem cell transplantation (SCT).[1–8] It is noteworthy that SMS is estimated to be represent in 0.7% of acute myeloid leukemia cases, while it may be concomitant in up to 8% of the patients with this leukemia recently diagnosed, and without treatment 71% of isolated tumors evolve to acute leukemia in less than one year.[5] Abedi and colleagues reported six representative case studies of this tumor type, including a 57-year-old man who had a subcutaneous tumor in the scalp for one month, without general symptoms, and presenting with normal results of the hemogram and bone marrow study.[1] Histopathological and immunohistochemistry evaluations of tumor samples revealed a granulocytic sarcoma with involvement of the subcutaneous fat; he underwent cytarabine and idarubicin to obtain a remission, besides the high-dose of cytarabine and radiotherapy. Two years later, there was a tumor recurrence in a subcutaneous area of the neck, and the patient received a similar chemotherapy schedule followed by the hematopoietic SCT.[1] Another 66-year-old man with manifestations indicative of sinus tumor, and diagnosis of granulocytic sarcoma by biopsy studies, presented normal hemogram and bone marrow. He was then uneventfully treated with AML induction chemotherapy and radiotherapy.[1] A 26-year-old female with manifestations of acute intestinal obstruction had abdominal imaging studies showing a mass of 5 cm and enlarged lymph nodes in the cecal region, which was surgically excised and accurate histopathological studies confirmed the diagnosis of myeloid sarcoma.[1] Bone marrow aspiration and hemogram were normal, and the patient underwent a routine schedule for AML induction chemotherapy.[1] A 53-year-old woman with a 6 years antecedent of AML remission had genital ulcers for two months, with the hypothesis of pyoderma gangrenosum resistant to treatment, and a new skin biopsy evaluation revealed the diagnosis of myeloid sarcoma (MS).[1] The data of hemogram and bone marrow aspiration were normal, and the genital ulcers healed by AML induction therapy.[1] A 32-year-old man had respiratory manifestations during two months associated with collateral vein circulation in the chest wall, and the imaging studies showed a large mediastinal mass compressing the esophagus, pulmonary arteries, and the vena cava.[1] The studies of tumor biopsy

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samples established the diagnosis of MS; while the results of hemogram and bone marrow aspiration and biopsy revealed unremarkable findings; he underwent the AML induction chemotherapy schedule with a clinical improvement.[1] A 72-year-old woman with AML on remission had cervical lymphadenopathy during a month, and biopsy samples revealed disarrayed structure and neoplastic cells infiltration, with histopathological and immunohistochemical characteristics consistent with MS.[1] As the aspiration and biopsy of bone marrow showed normal hematopoietic elements, the authors concluded that MS was a relapse of AML in a patient treated by chemotherapy.[1]

In this setting, it seems profitable to add some comments on novel references.[2–8] A 29-year-old male with dysuria and intense lumbar pain radiating to lower limbs during one month and unresponsive to clinical management; imaging studies showed an iso-dense soft mass compressing the spinal cord at L2 to L4.[2] Because of the progressive intensification of the clinical manifestations, he underwent the prompt resection of the tumor (6.0×2.5×1.2 cm) and the final diagnosis of MS was established.[2] As the repeated hemograms and the result of the bone marrow examination were normal, the case study was consistent with isolated MS without the bone marrow involvement.[2] The patient had additional chemotherapy with daunorubicin and cytosine arabinoside and evolved with clinical improvement during the follow-up, and without AML involvement.[2] A 7-month-old male infant presenting with partial paralysis of the lower limbs due to compression by a thoracic and intraspinal non-leukemic MS.[3] He also developed recent dyspnea and typical signs of right-sided pleural effusion (confirmed by images), containing monoblastic cells, with aberrant expression of CD7 and CD56. Besides, there was a tumor involving the right pleura, the chest wall, the mediastinum, and the neural foramina, in addition to the intra-abdominal development along the blood vessels.[3] The examination of bone marrow aspirate was entirely normal, and the diagnostic conclusion was consistent with the uncommon extramedullary manifestations of an isolated MS.[3] The protocol AML-BFM 2012 was utilized and the palsy reduced with cytarabine use; but 16% blasts were found in the bone marrow aspirate of control; the patient was re-stratified to high-risk group and allogeneic SCT indicated in the first complete remission.[3] After the fourth chemotherapy cycle, he underwent SCT, receiving peripheral blood stem cells (PBSC) from a matched unrelated donor, and after 17 months there was no residual evidence of the MS.[3] A 47-year-old male with lumbar spinal cord compression by a MS without bone marrow change or any hematological manifestation; he successfully underwent L4 and L5 laminectomy and debulking besides radiotherapy.[4] The hemogram, as well as the evaluation of the bone marrow biopsy samples were unremarkable. The preoperative imaging studies showed an intrathecal mass at L4–L5 with characteristics of schwannoma, but the histopathological data of samples indicated a round blue cell tumor, and the final diagnostic conclusion was for extramedullary manifestation of AML as MS.[4] Because of postoperative persistence of symptoms, in absence of bone marrow changes and locally infiltrating unresectable disease, he underwent a radiation therapy for relief.[4] The authors emphasized the rare occurrence of an isolated MS affecting the lumbar spine. A 76-year-old cirrhotic woman with a painful mass in the abdominal wall, multiple scattered bumps, and imaging studies revealing subcutaneous soft tissue lesions measuring 0.5 to 5.5 cm, in addition to bilateral pulmonary nodules.[5] The preliminary skin biopsy diagnosis was a "high-grade hematolymphoid neoplasm with differential including MS and a blastic plasmacytoid dendritic cell neoplasm"; but the conclusive pathology diagnosis including new biopsy studies of the abdominal mass was isolated MS, which was not treated because she presented skin abscesses with sepsis, and died.[5] The authors stressed the role of early biopsy studies to clear the challenging diagnosis. An 87-year-old woman with recent paraparesis due to an epidural tumor compressing the T4–T7 cord segments and the resected tumor was a MS.[6] After surgery, the strength in lower limbs improved, but the bone marrow aspirate established the diagnosis of a MS with monocytic differentiation, and the patient decided not underwent treatment by chemotherapy or radiotherapy, and died after four months.[6] The authors highlighted the rarity of the extra-axial MS, and the absence of consensual protocol to guide the specific surgical excision procedure, and the chemotherapy or radiotherapy for this malignancy.[6] A 25-year-old man with recent back pain who had initial biopsy diagnosis and treatment for tuberculous spondylitis without improvement, and presented with a gradually increased pain besides paraparesis in the lower limbs.[7] The imaging evaluations revealed pleural effusion, a T8 vertebral collapse, and an epidural collection which was the origin of a spinal cord compression from T7 to T9.[7] He underwent a T8–T9 laminectomy with epidural debridement, and in T8 vertebral body there was a grayish-white lesion but tuberculosis was discarded by specific investigation. With one month of postoperative course, he had dyspnea and cough and chest images showed osteolytic lesions in from T7 to 79 vertebral bodies, and in the left 7th rib that was biopsied and revealed the diagnosis of an extramedullary MS. The malignancy was promptly man-

aged by the cytarabine and daunorubicin schedule, which resulted in one year of with an unremarkable follow-up.[7] The authors highlighted the rarity of the spinal MS and the important role of accurate biopsy studies to confirm and differentiate this tumor from tuberculous spondylodiscitis.[7] A 50-year-old diabetic man who had numbness and weakness in lower extremities for a week, and imaging studies showed changes from T1 to T4 vertebral bodies, and a large lobulated extradural lesion within the spinal canal involving left neural foramina; besides a left paravertebral lesion extending to near the left apical pleura with contralateral displacement and focal buckling of the spinal cord.[8] The patient promptly underwent decompression of T1 to T3 vertebrae with complete excision of the lesion, and the histopathological evaluations established the consistent diagnosis of MS. Moreover, the induction chemotherapy was also indicated as component of the initial treatment plan.[8] The authors commented on the rare spinal involvement by the MS, which may propitiate a higher morbidity and disability in cases with tardive detection or late management.[8]

In conclusion, these short comments on recent literature aim to enhance the general interest among non-specialist health care workers about the main clues of MS diagnosis.

ΠΕΡΙΛΗΨΗ

Μυελοειδές σάρκωμα σε μη λευχαιμικούς και λευχαιμικούς ασθενείς

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Το μυελοειδές σάρκωμα είναι μια σπάνια οντότητα που μπορεί να αναπτυχθεί σε συνδυασμό με μια μυελοϋπερπλαστική νόσο, κυρίως την οξεία μυελογενή λευχαιμία, ενώ μπορεί να ανιχνευτεί πριν, ταυτόχρονα ή μετά τη διάγνωσή της. Αξίζει να σημειωθεί ο σπάνιος αριθμός επιβεβαιωμένων μεμονωμένων περιπτώσεων μυελοειδούς σαρκώματος. Αυτό το κακόηθες νεόπλασμα είναι επίσης γνωστό και ως κοκκιοκυτταρικό σάρκωμα, μυελοβλάστωμα ή χλώρωμα και έχει περιγραφεί στο δέρμα, στους λεμφαδένες, στο λεπτό έντερο και στα οστά, συχνά με αρκετά δύσκολη διαγνωστική προσέγγιση. Η πρώιμη εμφάνιση αυτού του όγκου στη βρεφική ηλικία είναι πολύ σπάνια και οφείλεται μάλλον σε γενετική προδιάθεση, ενώ μεταξύ των ενηλίκων η εκτιμώμενη επίπτωση του μεμονωμένου όγκου είναι 2 ανά 1 εκατομμύριο πληθυσμού. Δεν υπάρχει κάποιο συναινετικό πρωτόκολλο χειρισμών, αλλά οι επιλογές περιλαμβάνουν χειρουργική εκτομή, χημειοθεραπεία, ακτινοθεραπεία και μεταμόσχευση βλαστοκυττάρων. Στόχος είναι η αύξηση της ευαισθητοποίησης των εργαζομένων στον τομέα της υγείας σχετικά με τις κύριες διαγνωστικές ενδείξεις.

Λέξεις ευρετηρίου: Αντιμετώπιση, Διάγνωση, Λευχαιμία, Μυελοειδές σάρκωμα

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