2. Zanetti G, Nobre LF, Mançano AD, et al. Nodular reversed halo sign caused by pulmonary tuberculosis, confirmed by sputum culture. Radiol Bras. 2013;46(6):ix–x.
3. Amoedo MK, Souza LVS, Souza AS, et al. Pulmonary interstitial emphysema: a case report and review of the literature. Radiol Bras. 2013;46: 317–9.
4. Koenigkam Santos M, Barreto ARF, Chagas Neto FA, et al. Neuroendocrine tumors of the lung: major radiologic findings in a series of 22 histopathologically confirmed cases. Radiol Bras. 2012;45:191–7.
5. Zanetti G, Nobre LF, Mançano AD, et al. Paracoccidioidomicose pulmonar. Radiol Bras. 2014;47(1):xi–xiii.
6. Fernandes MC, Zanetti G, Hochhegger B, et al. Pneumonia por *Rhodococcus equi* em paciente com SIDA. Radiol Bras. 2014;47(3):xi–xiii.
7. Koenigkam-Santos M, Paula WD, Gompelmann D, et al. Endobronchial valves in severe emphysematous patients: CT evaluation of lung fissures completeness, treatment radiological response and quantitative emphysema analysis. Radiol Bras. 2013;46:15–22.
8. Hochhegger B, Felicetti JC, Marchiori E. An uncommon intrafissural mass: pleural primitive neuroectodermal tumor. Arch Bronconeumol. 2014 Mar 27. doi: 10.1016/j.arbres.2014.01.003. (Epub ahead of print].
9. Pandit S, Mukherjee S, Bhattacharya S, et al. A rare mediastinal tumour in a young male mimicking massive pleural effusion. Lung India. 2012;29:66–9.
10. Biswas B, Agarwal AS, Shukla NK, et al. Evaluation of outcome and prognostic factors in thoracic primitive neuroectodermal tumor: a study of 84 cases. Ann Thorac Surg. 2013;96:2006–14.
11. Parikh M, Samujh R, Kanojia RP, et al. Peripheral primitive neuroectodermal tumor of chest wall in childhood: clinico-pathological significance, management and literature review. Chang Gung Med J. 2011; 34:213–7.
12. Shi L, Guo Z, Wu X. Primary pulmonary primitive neuroectodermal tumor metastasis to the pancreas: a rare case with seven-year follow-up. Diagn Pathol. 2013;8:51.

Gilmar Moreira Silva Junior[1], Gláucia Maria Ribeiro Zanetti[2], Jorge Luiz Barillo[3], Edson Marchiori[2]

1. Lumic – Radiology, Petrópolis, RJ, Brasil. 2. Universidade Federal do Rio de Janeiro (UFRJ), Rio de Janeiro, RJ, Brasil. 3. Universidade Federal Fluminense (UFF), Niterói, RJ, Brasil. Mailing Address: Dr. Edson Marchiori. Rua Thomaz Cameron, 438, Valparaíso. Petrópolis, RJ, Brazil, 25685-120. E-mail: edmarchiori@gmail.com.

http://dx.doi.org/10.1590/0100-3984.2014.0019

Melorheostosis

Dear Editor,

A female, 27-year-old patient attended the health unit complaining of pain in her left upper limb for six months. The patient denied trauma or fever. Physical examination revealed increased limb volume, skin hardening, with no flogistic sign. Laboratory tests did not demonstrate any significant alteration.

Plain radiography of the affected limb and hip demonstrated alterations typical of melorheostosis, such as unilateral cortical hyperostosis along the bone axis resembling "melted candle wax" involving only one body segment (left hemibody) and extending from the humerus up to the hand bones (Figure 1). Similar alterations were also observed in the iliac bone at left (Figure 2)

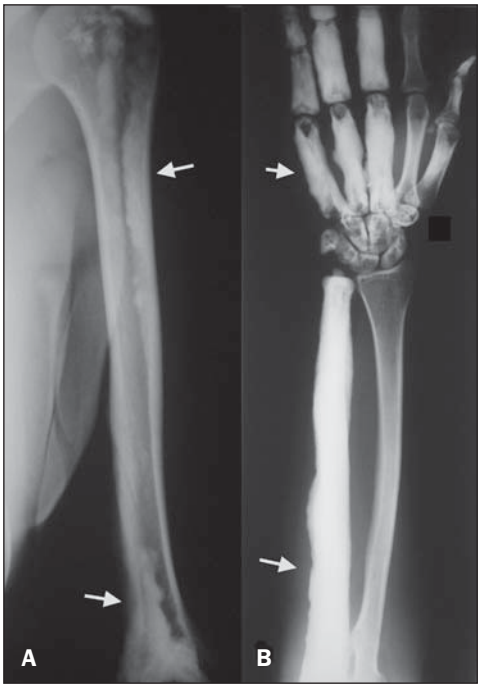

Figure 1. Radiographic images of left arm (**A**), forearm and hand (**B**) demonstrating cortical hyperostosis along the bone axis resembling "melted candle wax" (arrows).

Melorheostosis is a rare sclerosing disease of unknown etiology characterized by involvement of, particularly, long bones and adjacent soft tissues. It has a peculiar radiological characteristic, i.e. cortical thickening resembling "melted candle wax"[1–3]. Both female and male individuals are equally affected. The disease onset may occur at any age, but in 50% of cases it occurs before the age of 20[1].

Although it is a benign disease, it may bring clinically significant morbidity to patients. Initially it may be asymptomatic, but the association between bone sclerosis and tissue fibrosis leads to shortening of limbs, bone deformity and joints stiffness, gradually progressing to intense pain and functional incapacity of the affected limb.

Bone cortical thickening is the main pathological alteration, with both mature and immature components, thickened bone trabeculae and increased osteoblastic activity[1,2,4,5], that may extend to joints. It is common to observe involvement of adjacent soft tissues, with development of fibrotic, bone, cartilaginous and vascular tissues.

The disease distribution is peculiar and typically affects only one side of the body, and may be either mono- or polyostotic, in

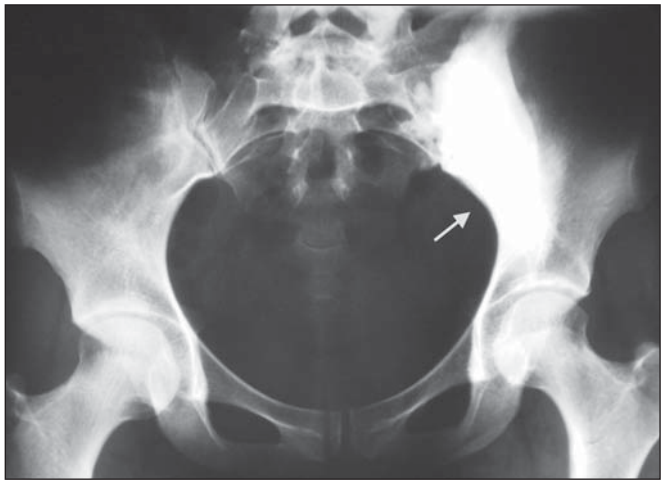

Figure 2. Radiographic image of hip shows iliac and sacrum hyperostosis (arrow), at left, with the same finding observed in the upper limb of the patient.

this latter presentation following sclerotomal distribution. Long bones of the lower limbs are most frequently affected[1–5]. The disease rarely affects the spine, skull and the face. The diagnosis is essentially clinical and radiological. Laboratory tests results are usually normal and histological findings are nonspecific.

The classical radiological presentation is that of sclerosis in only one side of the cortical bone, with linear and segmental distribution, resembling melted candle wax dropping along the bone axis and projecting over the medullary space. Such an alteration may distally extend to finger bones.

Other observed presentations are similar to osteoma, striated osteopathy, osteopoikilosis, and myositis ossificans, with calcifications in adjacent soft tissues [1].

Computed tomography (CT) shows in more detail the sclerotic alterations as well as reduction of the medullary space. At magnetic resonance imaging (MRI) such alterations present with low signal intensity at T1- and T2-weighted sequences, a finding that is compatible with cortical bone. The involvement of soft tissues may also be observed, with variable calcification degrees at CT; and MRI shows images with heterogeneous signal intensities corresponding to mineralization, areas of fat and fibrovascular tissue[1,2].

Thus, melorheostosis is highlighted as a relevant differential diagnosis among bone diseases, particularly because of the characteristic radiographic findings of this disease.

REFERENCES

1. Suresh S, Muthukumar T, Saifuddin A. Classical and unusual imaging appearances of melorheostosis. Clin Radiol. 2010;65:593–600.
2. Nuño C, Heili S, Alonso J, et al. Melorreostosis: presentación de un caso y revisión de la literatura. Rev Esp Enferm Metab Oseas. 2001;10:50–5.
3. Salman Monte TC, Rotés Sala D, Blanch Rubió J, et al. Melorheostosis, a case report. Reumatol Clin. 2011;7:346–8.
4. Mariaud-Schmidt RP, Bitar WE, Pérez-Lamero F, et al. Melorheostosis: unusual presentation in a girl. Clin Imaging. 2002;26:58–62.
5. Gagliardi GG, Mahan KT. Melorheostosis: a literature review and case report with surgical considerations. J Foot Ankle Surg. 2010;49:80–5.

Paulo Marcus Vianna Franca[1], Cid Sérgio Ferrreira[1], Reginaldo Figueiredo[1], João Paulo Kawaoka Matushita[1]

1. Universidade Federal de Minas Gerais (UFMG), Belo Horizonte, MG, Brasil. Mailing Address: Dr. Paulo Marcus Vianna Franca. Rua Itaí, 570, ap. 102, Santa Efigênia. Belo Horizonte, MG, Brazil, 30260-290. E-mail: pmviannafranca@gmail.com.

http://dx.doi.org/10.1590/0100-3984.2013.0019

Pre- and postnatal findings of a *dicephalus tetrabrachius-dipus* conjoined twins with a diaphragmatic hernia

Dear. Editor,

A 17-year-old primigravida attended the service at the 31st gestational week for evaluation of monochorionic, monoamniotic twin gestation. First trimester sonographic images were not available. Morphological ultrasonography (US) demonstrated the fetuses joined at the level of their abdomen and pelvis, and presence of a diaphragmatic hernia in the second twin. The woman denied previous history of health problems or use of medicines and illicit drugs. Her 25-year-old husband was healthy, with negative history of consanguinity. No family history of genetic diseases and malformations was reported. Fetal magnetic resonance imaging (MRI) revealed a *dicephalus tetrabrachius-dipus* conjoined twin. The fetus at right presented with a left diaphragmatic hernia containing stomach, small bowel and colon. The twins shared a single liver and a urinary bladder. Two kidneys connected each other at the level of their lower poles, and two vertebral spines were fused at the level of the sacrum (Figure 1). Echocardiography was normal.

The conjoined twin was born by Cesarean section at the 35th gestational week, weighting 3,765 grams. Upper eyelid coloboma was found in the twin with diaphragmatic hernia. Radiographic evaluation demonstrated vertebral spines fusion at the level of the lumbar region, besides the presence of bowel loops in the thoracic cavity of the twin at right (Figure 2). Surgery for the diaphragmatic hernia could not be performed. The conjoined twin died at the 17th day of life.

Imperfect twinning occurs in approximately one per 250,000 live births[1,2] and is classified according to the fusion site added by the term *pagus*[3]. "Parapagus" twins (meaning "extensive lateral fusion") correspond to 28% of cases of conjoined twins[4]. The subtype *dicephalus tetrabrachius-dipus*, as observed in the present case, is considered rare (4/10,000,000 births)[5].

US has shown to be the best method for initial evaluation of the gestation, and can identify imperfect twinning as early as at the 12th gestational week[1]. However, US is subjected to limitations such as maternal biotype and presence of either oligohydramnios or anhydramnios. On the other hand, MRI represents a good complementary tool since it does not present such limitations, while providing images with better resolution[6]. Additionally, it serves as support for a possible surgical planning, since it allows for visualization and detection of abnormalities which otherwise would be missed or inconclusive at US[2]. In the present case, MRI was relevant, particularly in the determination of the type of imperfect twinning as well as of the extent of fusion and sharing of organs.

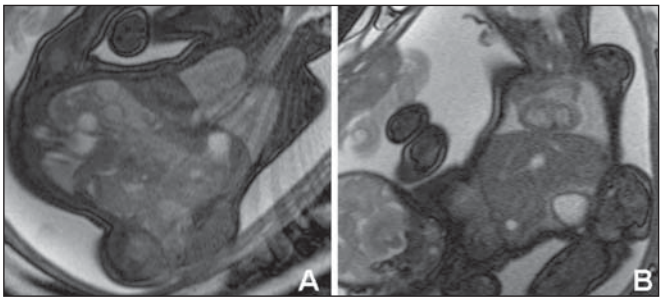

Figure 1. Fetal MRI T2-weighted image showing the *dicephalus tetrabrachius-dipus* conjoined twin. The fetus at right presents with a left diaphragmatic hernia. Mediastinal structures (heart and large vessels) and pulmonary hypoplasia (**A**) are identified. Hepatic fusion is also visualized (**B**).

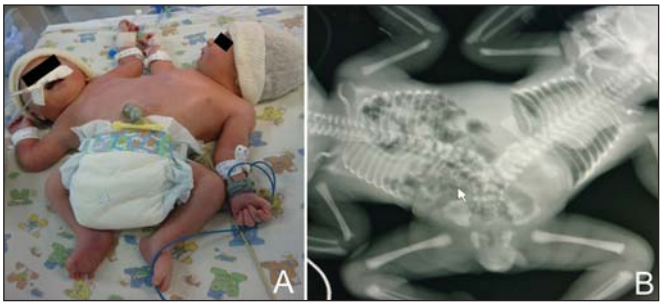

Figure 2. Postnatal image of the *dicephalus tetrabrachius-dipus twin* (**A**). Radiographic evaluation showing vertebral spines fusion at the L4 level and a single pelvis. A diaphragmatic hernia is observed in the fetus at right (there is evidence of the presence of bowel loops within the thoracic cavity), without identification of the heart and airways (**B**).