## Maladie osseuse de Paget

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Un homme de 65 ans a été adressé en clinique d'endocrinologie après avoir obtenu un taux élevé de phosphatase alcaline (PA), à 332 U/L (éventail des valeurs normales : 30–120), dans des analyses effectuées dans le cadre d'une visite périodique.

Le taux de PA osseuse spécifique était élevé, à 83.3 µg/L (éventail des valeurs normales : 5,6-29), tandis que les taux sériques de 25 hydroxyvitamine D, de γ glutamyl transférase, de calcium, de phosphore et de parathormone étaient normaux. L'homme n'a fait état d'aucun antécédent de fractures ou de douleurs osseuses, de perte auditive ou de céphalées. La radiographie ordinaire a montré des anomalies osseuses typiques de la maladie de Paget au fémur proximal droit (figure 1). La scintigraphie osseuse a, pour sa part, révélé une fixation accrue du traceur radioactif au tiers proximal du fémur droit, au sacrum, aux régions préorbitaires et à la vertébre T3 (annexe 1, accessible en anglais au www.cmaj.ca/lookup/doi/10.1503/cmaj.230164/tab-related-content). Un diagnostic de maladie osseuse de Paget a été posé, et une dose de 5 mg d'acide zolédronique a été administrée par voie intraveineuse. Six mois après le traitement, le taux de PA s'était normalisé, et l'état du patient était toujours asymptomatique.

La maladie de Paget est un trouble focal du métabolisme osseux. Sa prévalence augmente avec l'âge, et elle affecte de 1%-5% des personnes de plus de 50 ans<sup>3</sup>. La plupart des cas de maladie de Paget sont asymptomatiques; le symptôme qui amène le plus souvent les gens à consulter est la douleur osseuse. On soupçonnera la maladie de Paget devant un taux élevé de PA, et le diagnostic repose sur la présence d'anomalies caractéristiques à la radiographie, comme l'épaississement trabéculaire et cortical, une déformation osseuse et l'ostéosclérose. Il n'est pas nécessaire de procéder à un examen du squelette, mais une scintigraphie osseuse devrait être réalisée au départ afin de révéler l'étendue de la maladie<sup>1</sup>. Le traitement est indiqué dans les cas d'atteinte des os porteurs, en l'absence de symptômes dans le but de réduire le risque de fracture et de déformation de certains os, mais cette pratique s'appuie sur des données de qualité médiocre<sup>1</sup>. Chez les personnes qui n'ont pas de symptômes, chez qui l'atteinte osseuse est peu étendue et le taux de PA, légèrement élevé, on procédera à des vérifications biochimiques annuelles, sans traiter. Le traitement à privilégier est une seule perfusion intraveineuse de 5 mg d'acide zolédronique, que l'on peut répéter si le taux de PA ne revient pas à la normale<sup>a</sup>. Il n'est pas nécessaire de reprendre les épreuves d'imagerie, mais l'apparition de nouvelles lésions osseuses justifierait que l'on procède rapidement à des examens à la recherche d'autres troubles que la maladie de Paget.



Figure 1: Radiographie du bassin chez un homme de 65 ans, qui montre une structure trabéculaire grossière au fémur droit, accompagnée d'un épaississement cortical marqué (filéche) et d'un rétrécissement de l'espace articulaine qui témoignent d'une arthrose secondaire à une déformation pagitique.

#### Références

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Intérêts concurrents : Aucun déclané.

Cet article a été révisé par des pairs.

Les auteurs ont obtenu le consentement du patient.

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