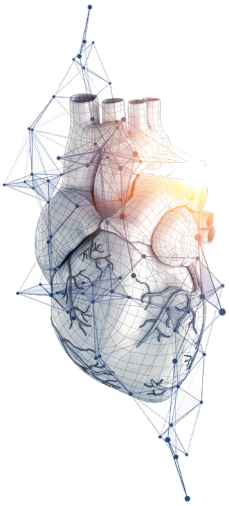

BÖLÜM 31

BİR OLGU ÜZERİNDEN KARDİYAK AMİLOİDOZA YAKLAŞIM

Zehra GÜVEN ÇETİN [1]

ÖZET

Amiloidoz, proteinlerin hatalı katlanması sonucunda dokularda birikmesi ile oluşan multisistemik bir hastalıktır. Kardiyak amiloidoz miyokardda ekstraselüler amiloid fibrillerinin birikimi ile seyreder ve sistemik amiloidozlar içinde prognozu en kötü olan tiptir. Daha önceleri nadir bir hastalık olarak kabul edilse de son yıllarda tanısal tekniklerin gelişmesi ile karşımıza daha sık çıkmaktadır. Kardiyak amiloidoz kalp yetmezliğinden ileti bozukluklarına kadar farklı klinik bulgular ile prezante olabilir. Bu yazıda sunulan olgumuz 85 yaşında kalp yetmezliği kliniği ile başvuran ekokardiyografide sol ventrikül hipertrofisi ve restriktif tipte doluş bozukluğu olması üzerine kardiyak amiloidoz şüphesi ile tektik edilen bir hastadır. Güncel tanı algoritmalarına göre girişimsel olmayan yöntemler ile tanı almıştır ve tedavi planı yapılmıştır. Bizim olgumuz gibi günlük pratiğimizde sık karşılaştığımız belirti ve bulgulara sahip olan hastalarda kardiyak amiloidoz akılda tutulması gereken bir hastalık olup tanıda en önemli aşama klinik şüphedir. Günümüzde yeni ve umut veren gelişmeler sayesinde amiloidoz artık tedavi şansı olan bir hastalık olarak kabul edilmektedir.

GİRİŞ

Kardiyak amiloidoz son yıllarda farkındalığı ve popüleritesi artmasına rağmen klinik pratikte halen nadiren tanı konulan bir hastalıktır. En sık korunmuş ejeksiyon fraksiyonlu kalp yetmezliği kliniği ile karşımıza çıkmaktadır.

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Anti TTR antikorlar: ATTR kardiyak amiloidoz tedavisinde immünoterapi kategorisinde incelenen ilk ilaç anti serum amiloid P komponent antikoru ile yapılan faz 1 çalışmasında amiloid fibrillerin eliminasyonu açısından umut verici sonuçlar alınmasına rağmen faz 2 çalışması erken sonlandırılmıştır[52]. Prothena (PRX004) ve NI006 ATTR tedavisinde çalışmaları devam eden iki yeni immünoteröpatik ajandır.

SONUÇ

Kardiyak amiloidoz progresif ve mortalitesi yüksek bir hastalıktır. Amiloidozun organ tutulumları içinde en kötü prognoza sahip olanıdır. Son yıllarda klinik pratikte daha sık tanınmasına rağmen halen tanı atlanmaktadır. Tanıda en önemli nokta klinik olarak şüphe duymaktır. Tanısal yöntemlerde meydana gelen gelişmeler sayesinde kardiyak amiloidozu görüntülemek daha kolaylaşmıştır ve biyopsi gerekliliği azalmıştır. Amiloidoz alanındaki diğer önemli gelişmeler ise tedavi alanındadır ve amiloid spesifik tedaviler ile amiloid birikimini azaltıp/durdurup yaptığı organ hasarlarını geri döndürmek hedeflenmektedir. Klinik çalışmaların sonuçları umut vaat edicidir. Devam etmekte olan randomize kontrollü çalışmaların sonuçları da güncel tedavilere yön verecektir.

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