Mortality And Frequency Of Nephrogenic Systemic Fibrosis

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Introduction:
Nephrogenic Systemic Fibrosis (NSF) is a rare, debilitating condition affecting individuals with severe renal impairment. It associates with the administration of gadolinium-based contrast agents (GBCA) used for magnetic resonance angiography (MRA). Mortality is high in reported case series but little information exists on mortality compared to a matched population.

Methods:
1. The electronic patient records (EPR) for two Glasgow renal units were searched to identify all patients on renal replacement therapy (RRT) from 01/01/2000 to 01/07/2006. Those with a functioning transplant and acute kidney injury were excluded. NSF and GBCA exposure were identified using EPR textfinder and radiology screen searches, case note review, dermatology and pathology contact, eCRIS searches and biopsy review. Onset of RRT and outcome including date of death was recorded. Survival to death or census (01/07/06) was determined from either onset of RRT or scan date and analysed using SPSS software.
2. To identify if changing patterns of MRA referral have impacted on NSF case frequency, numbers and date of Glasgow wide referrals for all types of MRA from either ‘all’ or ‘renal’ referrers was recorded together with the date of onset of NSF cases.

Results:
1. 1826 patients were identified as having RRT within the 6.5 year study period. 425 received GBCAs and 14 developed NSF, 13 following gadodiamide exposure. 1812 had outcome data. 704 (38.9%) patients died and median survival from RRT onset to death or census for the whole cohort was 2.6 years. Median survival for the GBCA exposed cohort was also 2.6 years and median survival from scan to death or census was 2.0 years. Median survival for the gadodiamide exposed NSF population was not significantly different measured by RRT onset or post scan survival. No significant difference could be found when survival for the total RRT cohort minus gadodiamide exposed cohort was compared to the NSF population.
2. In total 17 cases of NSF are known to our unit presenting between January 2001 and January 2007. 16 cases were associated with gadodiamide. No further cases have been reported despite increased awareness. Between 1998 and 2002 MRA requests from renal referrers steadily increased year on year from 13 to 113. They remained steady and then declined following the link with GBCA and NSF to 32 in 2008. The frequency of NSF cases follows the frequency of MRA requests by renal referrers.

Conclusions:
1. Despite poor survival, no significant difference in mortality could be demonstrated between our NSF patients and an RRT population also selected for GBCA exposure.
2. The frequency of NSF cases closely follows the number of MRA scans requested by renal referrers suggesting that GBCA exposure is causally implicated in the development of NSF.